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VOL. 36 SEPTEMBER 1951 FASC. 3

SUPRASELLAR TUMOURS WITH CALCIFICATION[1]

by

Erik Lindgren and *Giovanni Di Chiro*

Craniopharyngiomas

A correct localizing diagnosis of a suprasellar lesion is generally easy. Primary optic atrophy, visual-field distortions, sellar changes, suprasellar calcification, oculomotor palsies, signs of dyspituitarism and so on, taken together constitute a symptom-complex pointing unmistakably to the suprasellar region. On the other hand, the preoperative diagnosis of the morbid nature of the process is far more difficult. An analysis undertaken at CUSHING's suggestion by DEERY (12) in 1930 revealed that although the localizing diagnosis had been made preoperatively in all the 170 cases of suprasellar lesions reviewed, the morbid nature of the process had been correctly diagnosed in only 85 instances (exactly 50 per cent of the cases). The incorrect diagnosis had often been caused by a misinterpretation of the roentgen findings.

The appearance of the sella turcica and the presence or the absence of suprasellar calcification are the two main roentgenological features in studying suprasellar lesions. The finding of suprasellar calcification is a sign which is often considered to constitute a clue in the differential diagnosis. Suprasellar calcification means craniopharyngioma for most of authors; some, however, have found suprasellar calcification in other lesions besides craniopharyngiomas, but no systematic study of a large material has been carried out. The present writers therefore have con-

[1] Submitted for publication, June 5, 1951.

From the Roentgen Department (Director: Professor ERIK LINDGREN) of the Serafimerlasarettet, Stockholm, Sweden.

sidered it useful to review the suprasellar space-occupying lesions from the roentgenological standpoint with particular regard to the occurrence of calcification and the possibilities of making an accurate diagnosis. The material has been gathered in the roentgen department of the Serafimerlasarett. All the cases included have been pathologically verified.

The presence of lime salts within tumours arising from rests of Rathke's pouch was histologically demonstrated by ERDHEIM (17) as early as 1904. The term craniopharyngioma, coined by CUSHING, has been criticized (GLOBUS and GANG (19)) as incorrect from the embryological standpoint. Nevertheless the term is now well known and understood and it would be unnecessarily confusing to introduce a substitute. In 1909 ALGYOGI (2) described a case, which he considered as a tumour of the hypophyseal duct, presenting roentgenographically demonstrable calcification. The case was not pathologically verified. In 1914 LUGER (26) reported 3 cases of craniopharyngiomas (1 pathologically verified), in which suprasellar calcification had been roentgenographically demonstrated. JACKSON (25) in 1916 was able to collect in the literature 37 cases of craniopharyngiomas and added one personal case. He states: »The sella is rarely enlarged, and roentgenoscopy will usually be negative.» One must note that JACKSON's series is mainly based on post-mortem material. DUFFY (15) in 1920 added to JACKSON's series a further 17 cases (3 personal and 14 gathered from a more careful perusal of the literature), thus bringing the number of authentic cases reported at that time to a total of 55. DUFFY is far more optimistic regarding the value of roentgen findings in such tumours. The first important roentgenological work based on a large number of pathologically verified craniopharyngiomas is that of MCKENZIE and SOSMAN (28) (1924). These authors were able to detect suprasellar calcification in 25 (71 per cent) of the 35 cases reviewed (CUSHING's material).

Thereafter the literature on the subject becomes extensive and many authors begin to regard suprasellar calcification as the most important sign of tumours arising from the craniopharyngeal pouch. BÉCLÈRE (3) states: ». . . les taches suprasellaires ont pour le diagnostic des tumeurs de la poche de Rathke une valeur inestimable; elles sont vraiment pathognomiques.» VAN BOGAERT (36): »La signature pathognomique des tumeurs de la poche de Rathke est la présence de taches de calcification au dessus d'une selle turcique élargie ou normale.» VOGT (37): »L'aspect radiographique de ces tumeurs permet un diagnostic facile à cause des concrétions calcaires suprasellaires.» OLIVECRONA and LYSHOLM (31) referring to such calcification state: ». . . so gut wie pathognomisch ist» (of craniopharyngiomas). ZEITLIN (38): ». . . it may be considered pathognomic.» INGRAHAM and SCOTT (24): »This invaluable finding is almost pathognomic of the tumor in children.» There are, however, authors

(DEERY (12), SØRENSEN (35)) who make reservations, pointing out that suprasellar calcification is found also in tumours of other types besides craniopharyngiomas.

A high incidence of roentgenographically demonstrable calcification in craniopharyngiomas has been reported in the literature. MCKENZIE and SOSMAN (28) found calcification in 71 per cent of cases, other authors giving yet higher figures: DEERY (12) 72 per cent, DROGUET (14) 75 per cent, PUECH and STUHL (32) 75 per cent, CUSHING (7) 80 per cent, and DEW (13) 80 per cent of cases. The incidence of the finding of suprasellar calcification depends very much on the roentgen technique employed. The films must be sufficiently exposed. The suggestion given in 1924 by MCKENZIE and SOSMAN of a moderate overexposure of the films, probably finds its explanation in the fact that at that time the kilovoltage generally used for skull films was too low. High-definition intensifying screens and a focal spot of small effective size should be employed in order to obtain an optimum of sharpness. Some authors (VOGT, DROGUET) prefer stereoradiograms. As with other roentgenological examinations we think that the demonstration of suprasellar calcification by means of lateral and antero-posterior views is more objective. The most suitable a. p. view is taken with the tube angled 30/35 degrees cranially (what we call the halfaxial view in the supine position). In some cases satisfactory information may be obtained with a p. a. view (patient prone and tube angled 10—12 degrees cranially). Eroded clinoid processes and meningeal calcification may sometimes simulate calcification within a suprasellar lesion. The mistake can generally be avoided by a careful study of the appearance and position of the calcification. In these cases oblique views of the pituitary fossa region are useful. Nevertheless the review of our series of craniopharyngiomas gives a considerably lower incidence of suprasellar calcification than that reported by the above-mentioned authors. We have found suprasellar calcification in 30 of 54 verified cases of craniopharyngiomas — *i. e.* about 55 per cent. In order to clarify this point we have collected from the literature 238 cases of roentgen-examined and pathologically verified craniopharyngiomas. We have not attempted to exhaust the literature and we are aware that in our search a number of cases has been overlooked.

Of these 238 cases 139 showed suprasellar calcification — 58 per cent. In comparison to what is generally stated this is rather a surprising figure which, however, agrees closely with the 55 per cent found in our material.

Concerning the appearance of the calcification in question all the authors are in agreement with MCKENZIE and SOSMAN who give the following description: ». . . delicate and spongy in appearance, irregular in formation and not outlining a section of the cyst wall as in intra-

cranial aneurysms and dermoids.» PUECH and STUHL state that sometimes when the cystic content is particularly dense it is possible to see the complete cyst as a rounded, homogeneous structure within or above the sella. We have not seen such a case. Neither is our experience in agreement with the description of MCKENZIE and SOSMAN. In 19 of our cases the calcification has an appearance which we name »nodular» or »patchy» *i. e.* small spots rather sporadically and irregularly arranged within a certain area. On the other hand in 7 cases the calcification clearly outlines a section of the cyst wall. In these cases we speak of »cystic» calcification. In 4 cases the calcification is partly »nodular» and partly »cystic». We speak in such cases of »the mixed type». It is well known that the majority of the craniopharyngiomas are made up of a solid portion (generally the lower part) and of a preponderant cystic portion (generally the upper part). The cystic portion is more often constituted by several cysts of different size, and less often by a large alone cyst. In addition there are craniopharyngiomas which are totally solid. The operative prognosis is poor for all the craniopharyngiomas (CUSHING (9), GRANT (20)) but seems to be at its worst for the solid variety. It would therefore be useful for the neurosurgeon to know whether the tumour be cystic or solid. GUILLAUMONT and GUIOT (21) have suggested and successfully carried out the diagnostic and therapeutic puncture of cystic craniopharyngiomas in 7 cases by means of a needle inserted through the frontal bone and make the differentiation between cystic and solid craniopharyngiomas upon the presence of pyramidal signs and signs of raised intracranial pressure. They state: »Lorsqu'il s'y surajoute des signes d'hypertension intracranienne et des signes pyramidaux, on peut être sûr qu'il existe un kyste associé.» It is difficult to see why, as they suggest, a solid space-occupying lesion causes no increase of intracranial pressure and no signs of pyramidal tract involvement while a cystic lesion does so. In our opinion the appearance of the calcification may give information concerning the constitution of the craniopharyngiomas. In all our 7 cases in which the appearance of the calcification was of the »cystic» type, a solitary cyst, generally large, was found at operation. On the other hand, of the 4 solid tumours found at operation none had showed »cystic» calcification on roentgen examination. In the 4 cases in which the calcification was of »the mixed type» the operative finding was either of a tumour partly cystic and partly solid, or of a cystic tumour with solid incrustations on the walls. In the 19 cases in which the calcification was »nodular» the operation findings varied. In the most cases the tumour was partly cystic and partly solid with the calcification lying in the solid portion although in some cases the tumour was cystic with irregular calcified incrustations on the walls. In 2 cases the tumour was solid. Thus according to our experience »cystic» calcifica-

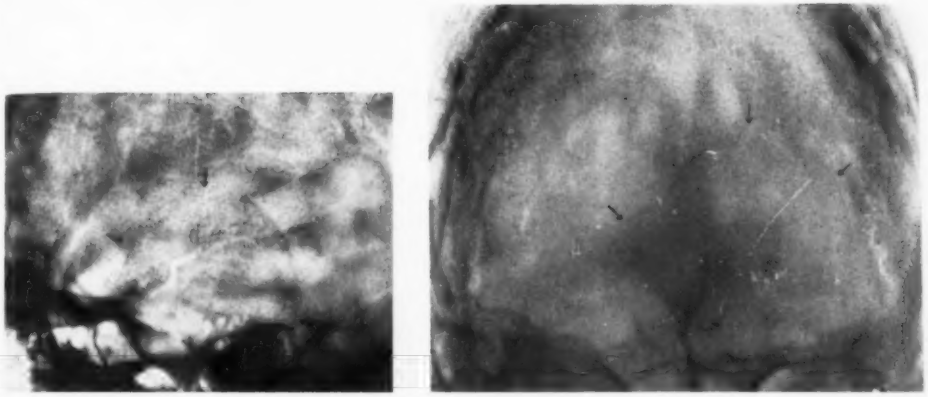


Fig. 1 a. Fig. 1 b.

Fig. 1. Craniopharyngioma. Large solitary cyst. Calcification outlines the entire cystic wall. »Flattened» sella. a) lateral view b) a.-p. (half-axial) view.

tion is indicative of a cystic tumour and calcification of »the mixed type» points to an at least partially cystic tumour. On the other hand when the calcification is »nodular» it is impossible to determine the constitution of the tumour. In such cases it may be expected to be partly cystic and partly solid although completely solid or completely cystic tumours may also occur.

The site of the calcification in our series is above the sella in 20 cases, both above and within the sella in 8 cases and only within the sella in 2 cases. With regard to the area, the »nodular» calcification has in our material varied in size from 2 × 2 mm to 4.5 × 4.5 cm as shown in the lateral view of the skull. Calcification of the »cystic» type has varied from one to several centimetres in length.

The sella in our series deviates from the normal in 47 instances (87 per cent of the cases). Among the 238 cases collected from the literature in which information about the sella is reported, 173 (72 per cent) showed an altered sella. The difference between our figures and those obtained from the literature may possibly be explained by individual variations in the interpretation of what constitutes an abnormal sella. We distinguish 4 different types of sellar changes. The first type we call »increased intracranial pressure sella» or more shortly »pressure sella» as it occurs in cases of raised intracranial pressure. The sella is enlarged and the dorsum and floor are more or less markedly destroyed. The earliest change consists solely of a disappearance of the corticalis of the floor and the anterior aspect of the dorsum, commonly at the junction

Fig. 2. Craniopharyngioma. Large solitary cyst. Calcification outlines the upper section of the cystic wall. »Cup-shaped» sella.

of these two formations. Changes of this first type are present in our material in 20 cases. The second type is the »cup-shaped» sella as in the chromophobe adenomas. Seventeen of our cases show this sellar shape. The third type is the »ballooned» sella as in the eosinophile adenomas. This type of sellar change is rare in craniopharyngiomas and when it occurs one may argue the exclusive intrasellar extension of the tumour. Three of our cases show this sellar shape. The fourth type is the most difficult to interpret — the dorsum is pushed downwards and is shorter than usual, the corticalis however being intact. This type of sellar configuration which we call »flattened sella» is present in 7 of our cases. In the remaining 7 cases the sella is normal.

The type of sellar changes occurring in craniopharyngiomas depends on two factors, namely the site of the epithelial remnants which have given origin to the newgrowth and the stage of development of the tumour. The schematic drawing of Fig. 5 shows the chief sites in which epithelial rests of the craniopharyngeal pouch are found. If a craniopharyngioma has originated from the lower group (L. G.) or the subhypophyseal group (S. G.) of epithelial cells, we get probably either the »cupshaped» or the »ballooned» sella. If the tumour has originated from the upper group (U. G.) of epithelial cells the sellar changes are usually of the first or of the fourth type, although occasionally of the second type. Operation has never shown an underdiaphragmatical development of the tumour in those of our cases in which the sella was of the »increased intracranial pressure» or of the »flattened» type. In 5 of the 17 cases in

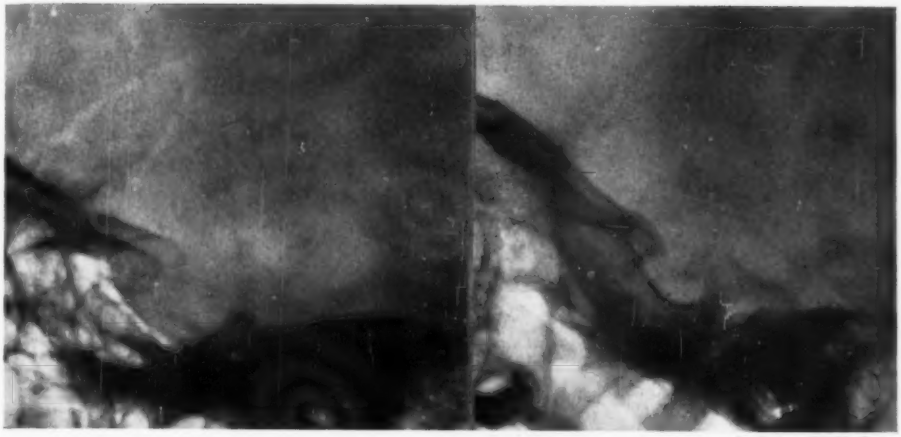


Fig. 3 a. Fig. 3 b.

Fig. 3. Craniopharyngiomas. Two examples of »nodular» calcification. Enlarged sella with destruction of the corticalis of the floor and the dorsum.
Operation showed in each case a tumour consisting of several cysts and an inferior solid portion.

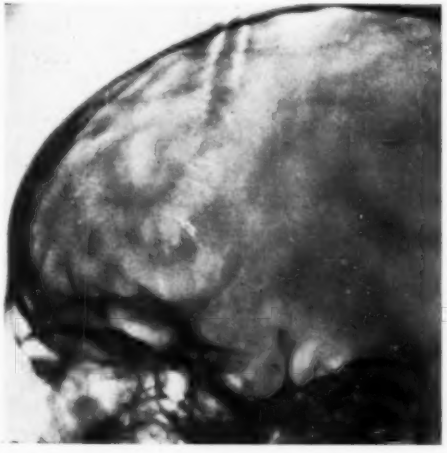


Fig. 4. Craniopharyngioma. »Mixed» *i. e.* part »cystic» and part »nodular» calcification. »Cup-shaped» sella and separation of the cranial sutures.
At operation a solitary cyst with calcified incrustations of the posterior wall was found.

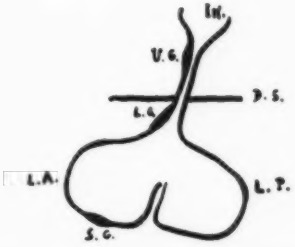
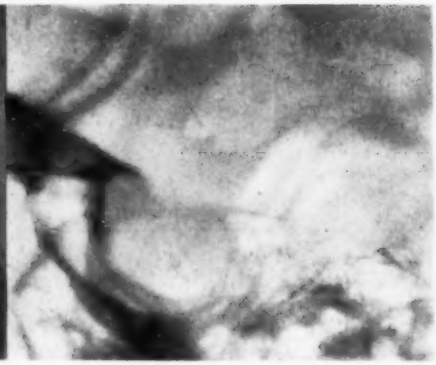


Fig. 5. Schematic drawing illustrating the chief sites in which epithelial rests of the craniopharyngeal pouch are found.
In. = infundibulum; D. S. = diaphragma sellae; L. A. = lobus anterior; L. P. = lobus posterior; U. G. = upper group of epithelial cells; L. G. = lower group of epithelial cells; S. G. = subhypophyseal group of epithelial cells.

Fig. 6 a.



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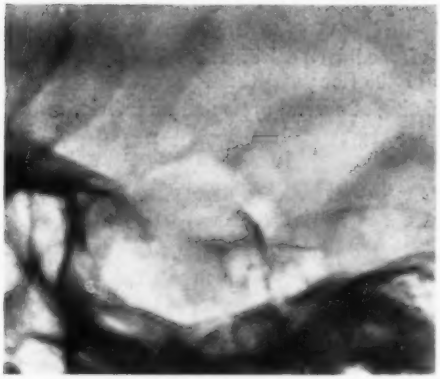


Fig. 6 c.

Fig. 6. Craniopharyngioma.

a) Preliminary film. »Nodular» intrasellar calcification. »Ballooned» sella.
b) Ventriculography. Anterior part of the third ventricle normal.
c) Encephalography. Anterior part of the third ventricle normal but the suprasellar cisterns are lifted up by an expansive lesion which has mainly an intrasellar extension.

which the sella was »cup-shaped» and in 1 of the 3 cases in which the sella was »ballooned», the operative records speak explicitly of underdiaphragmatical extension.

Other signs of increased intracranial pressure besides the sellar changes are not frequent in our series. Increase of the convolutional markings is present in 7 cases and separation of the cranial sutures in 10 cases. Definite widening of the chiasmal sulcus is present in 3 cases of our series.

McKenzie and Sosman and later Conley (6) described a depression of the sellar floor in the sphenoidal sinus with consequent more or less obliteration of the air cavity as being almost peculiar to craniopharyngiomas. This change has been interpreted by Cushing as due to a maldevelopment of the sphenoid bone associated with these congenital tumours. In our opinion the described change of the sphenoidal sinus is not characteristic of craniopharyngiomas but it may be found as frequently in other tumours of the pituitary region.

Fig. 7. Schematic drawing showing the deformation of the third ventricle and the disposition of the parasellar cisterns in craniopharyngiomas of fair size. The anterior part of the third ventricle is pushed slightly upwards and backwards. The tumour bulges into the anterior-inferior contour of the third ventricle between the anterior recesses. The parasellar cisterns surround the middle portion of the tumour.

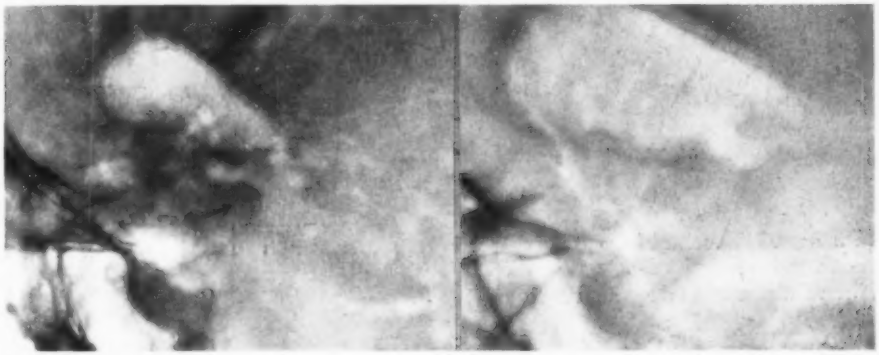


Fig. 8. Craniopharyngioma. Encephalography. Bulging of the tumour into the anterior-inferior part of the third ventricle. The two anterior recesses are conserved. Inferiorly the suprasellar cisterns delimit the neoplasm.

Operation showed a rather small completely suprasellar tumour.

Fig. 9. Craniopharyngioma. Encephalography. Only a part of the third ventricle near the Monroe's foramen is air-filled. The parasellar cisterns are pushed against the sellar entrance. In this and in similar cases a differential diagnosis of suprasellar gliomas is impossible by encephalography.

Reports of pneumographic studies dealing with craniopharyngiomas are scanty (Globus and Gang (19), Van Bogaert (36), Vogt (37)). One must note that 'air studies' carried out with deficient technique give only poor information in cases of craniopharyngiomas, as in other instances. As a rule symmetrical enlargement of the lateral ventricles has been found. Sometimes dilatation of the third ventricle has also been encountered, but authors generally agree that the third ventricle either does not fill at all or shows a filling defect.

In 22 of our cases encephalography has been performed and in 10 cases ventriculography carried out. The lateral ventricles are as a rule enlarged, the enlargement being sometimes of small degree and sometimes marked. The commonest change in the third ventricle consists of a slight displacement upwards and backwards, together with a filling

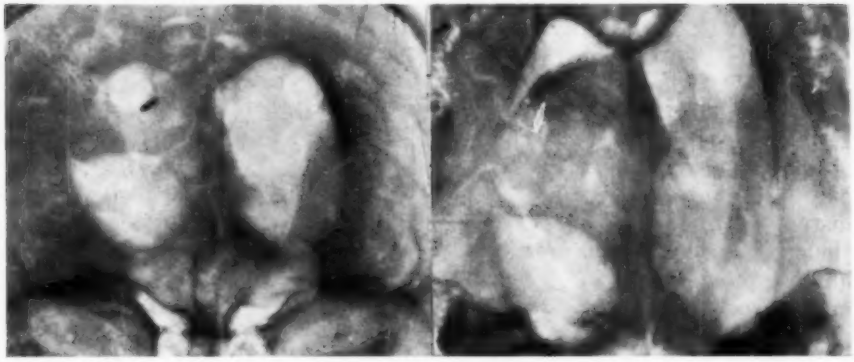


Fig. 10 a. Fig. 10 b.

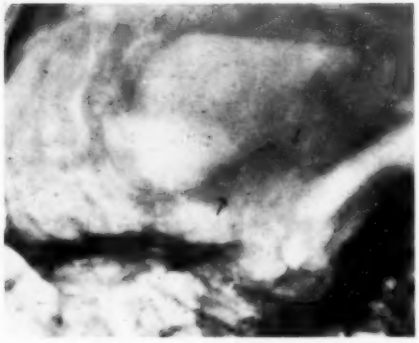


Fig. 10 c.

Fig. 10. Craniopharyngioma with »nodular» calcification (arrow). Ventriculography shows a large tumour which extends upwards into the lateral ventricles and occupies the main part of the right lateral ventricle. The picture resembles that found in some central gliomas.

defect which intervenes between the two anterior recesses (chiasmal recess and infundibular recess) but without obliterating them (Figs. 7, 8). If the tumour be large the compression and the displacement of the anterior part of the third ventricle is of such a degree that it is possible to fill only a small part of the third ventricle near the foramen of Monroe with air. Occasionally the third ventricle is not altered at all and the ventricular system is quite normal. An examination of the suprasellar cisterns is indispensable in order to assess exactly the extension of the tumour and its position in respect to the sella, especially in those cases in which the sellar changes are not clearly indicative and where the ventricular system is unaltered. If the tumour has solely suprasellar extension the suprasellar cisterns are pushed against the diaphragma sellae. On the other hand if the tumour has both supra- and intra-sellar extension the suprasellar cisterns are forcibly displaced and more or less completely surround the upper part of the neoplasm. In our series of 22 encephalographies combined supra- and intra-sellar extension of the tumour was seen in 10 cases, and in another 10 cases the exclusive suprasellar extension of the tumour was demonstrated.

Suprasellar Gliomas

In this group we include optic chiasma gliomas and hypothalamus gliomas. The differential diagnosis between suprasellar gliomas and craniopharyngeal pouch new-growths is extremely difficult. DEERY (12), who has reviewed 14 cases of optic chiasma gliomas reports that in 12 instances the diagnosis of craniopharyngioma had been made. Nevertheless it is commonly stated in the literature that the optic chiasma gliomas do not show calcification. MARTIN and CUSHING (27) suggest using suprasellar calcification as a differentiating sign between optic chiasma gliomas and craniopharyngiomas. In fact in none of the 7 cases of optic chiasma gliomas collected by these authors in 1923 was there any calcification. VAN BOGAERT, in 1929 referring to optic chiasma gliomas stated: ». . . on ne décèle jamais d'ombre calcaire suprasellaire.» Amongst the cases of optic chiasma gliomas which we have observed in the literature only two are reported as showing suprasellar calcification, namely, a case reported by HEUER and DANDY (23) and one of the 18 cases of the CUSHING's (8) new series of 1929. In 1934 HAMBY (22) reported a case of spongioblastoma bipolare in the region of the hypothalamus showing suprasellar calcification.

Our experience with 9 cases of optic chiasma gliomas and 5 cases of hypothalamus gliomas (all verified) is quite different. In 7 of these 14 cases (50 per cent) there is calcification, of optic chiasma gliomas in 3 cases and of hypothalamus gliomas in 4. The calcification lies in the suprasellar region in all the cases and has always the appearance which we have called »nodular» in the description of the roentgenological features of the craniopharyngiomas. The calcified area as shown in the lateral view of the skull varies from 1.5×1.5 cm to 3×2 cm. Possibly the elements constituting the calcification of suprasellar gliomas are of a smaller size and are a little more scattered in arrangement than in craniopharyngiomas but the appearance of the calcification is of little practical help in making a differential diagnosis. Among the 14 cases the sella shows deviations from the normal configuration in 11. Specifically the sella is of the »increased intracranial pressure» type in 8 cases, »flattened» in 2 cases and »cup-shaped» in 1.

In 6 cases there is increase of the convolutional markings and in 9 separation of the cranial sutures.

According to MARTIN and CUSHING (27) there is a roentgenological sign which may be important for the diagnosis of optic chiasma gliomas. The peculiarity is so described: ». . . there is a recognizable anterior extension of the fossa which passes like the neck of a gourd under the anterior clinoid processes. This is demarcated from the sella proper by the

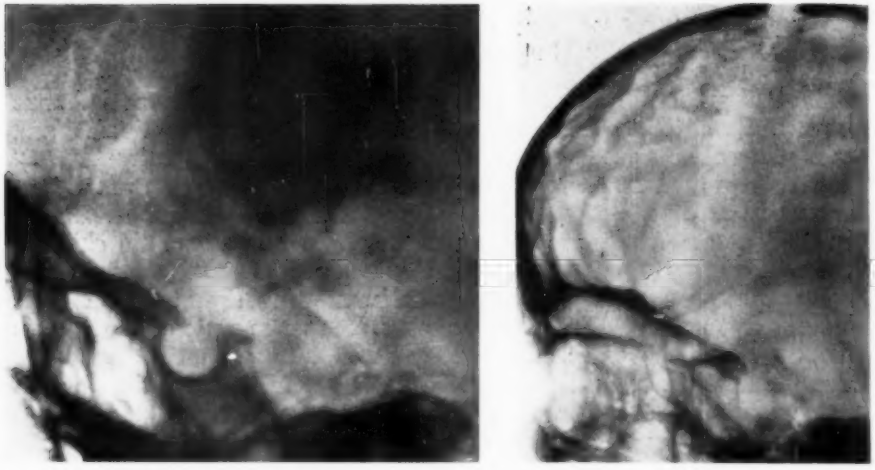


Fig. 11 a. Fig. 11 b.

Fig. 11. Optic chiasma gliomas with »nodular» calcification. In a) sella practically normal; chiasmal sulcus rather wide. In b) destruction of the corticalis of the floor of the sella and of the anterior aspect of the dorsum sellae; the chiasmal sulcus concave upwards gives a configuration to the sella which resembles an »hour-glass shape». Increase of the convolutional markings and separation of the cranial sutures.

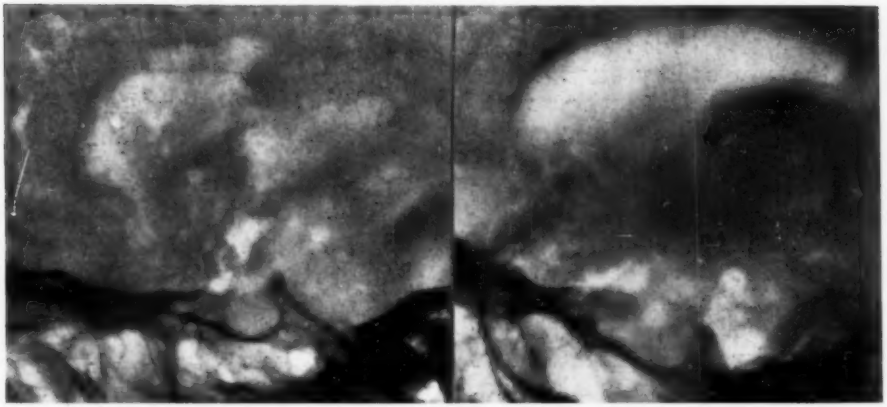
elevation made by the olivary eminence or tuberculum sellae. It is more marked in the cases with fully developed bilateral tumours and appears to be due to a pressure enlargement of the optic foramina.» This peculiarity gives to the sella a configuration described as »J-shaped» (American authors), »omega-shaped» (French authors), »hour-glass shaped» (German authors). The change is recognizable in the lateral view of the skull. None of our 9 cases of optic chiasma gliomas shows such a change. In one case however the chiasmal sulcus is enlarged and curved with the concavity upwards, the sellar configuration suggesting an »omega».

In 6 of the 9 cases of optic chiasma gliomas in our series, the optic foramina were examined with suitable projections. In 2 instances there was a definite bilateral enlargement of the foramina. Such enlargements are found however in pituitary adenomas and in other lesions in the region of the pituitary fossa.

Thus, in our experience, the plain roentgenograms of the skull give little information when there is a question of differentiating the suprasellar gliomas showing calcification from the craniopharyngiomas with »nodular» calcification. In fact the appearance of the calcification is in this respect of no value. Of relative value is the widening of the optic

foramina, possibly more frequent in the optic chiasma gliomas than in the craniopharyngiomas. The same must be said with regard to the increase of the convolutional markings and the separation of the cranial sutures, signs more frequently seen in the suprasellar gliomas than in the craniopharyngiomas. Too rare to be of practical differential importance is the anterior extension of the fossa under the anterior clinoid processes first described by MARTIN and CUSHING. The width of the chiasmal sulcus is also not of importance; it shows wide variations even in normal cases. On the other hand the signs noted in pneumography are of greater value in the differential diagnosis. The changes in the third ventricle occurring in suprasellar gliomas are of another type than those seen in craniopharyngiomas. The suprasellar gliomas push the third ventricle markedly backwards and sometimes bulge into its anterior border. In suprasellar gliomas we have on the other hand never seen, as in craniopharyngiomas, a bulging into the inferior contour of the third ventricle between the two anterior recesses; this contour is on the contrary often straightened and the two recesses compressed from in front (Figs. 12, 13). The difference in the change in the third ventricle found in craniopharyngiomas and in suprasellar gliomas may be explained by the different direction of the line of force. When the tumour has attained a large size, however, the third ventricle becomes nearly obliterated in both types of tumour and it is then impossible to obtain differential signs from the changes in the ventricular system. Even in cases of suprasellar gliomas the study of the suprasellar cisterns is useful for a complete elucidation of the anatomical conditions. While in craniopharyngiomas (and in pituitary adenomas) situated partly above and partly within the sella, the parasellar cisterns surround the upper part of the tumour, this never happens in suprasellar gliomas. It is usually difficult and often impossible to fill more than the posterior part of the suprasellar cisterns with air in a case of a suprasellar glioma; as a rule it is impossible to fill the cisterna laminae terminalis. In cases in which a filling of the cisterna chiasmatis with air is obtained the airfilled portion is pushed downwards. Thus a careful study of the anterior part of the third ventricle and of the suprasellar cisterns is rich in information in the differential diagnosis between

Fig. 12. Optic chiasma glioma. Ventriculography. Bulging of the tumour into the anterior contour of the third ventricle.



a) optic chiasma glioma.

b) schematic drawing.

c) hypothalamus glioma.

d) schematic drawing.

Fig. 13. Two examples of changes in the anterior part of the third ventricle and of the parasellar cisterns in cases of suprasellar gliomas of fair size. The third ventricle is pushed backwards and slightly upwards; the two anterior recesses of the third ventricle are not recognizable; the parasellar cisterns are only partially air-filled in a) and pushed apart in c). (Compare Figs. 7, 8.)

suprasellar gliomas with calcification and craniopharyngiomas presenting »nodular» calcification.

Other Lesions with Suprasellar Calcification

The two above mentioned groups of tumours represent the conditions in which suprasellar calcification is most frequently found. Several other lesions in the sellar region may however present such calcification. DEERY (11) in a review of 285 verified *pituitary adenomas* (CUSHING's series) found calcification within or just above the sella in 19 cases (6 per cent). In our material of 345 cases we have found calcification in 11

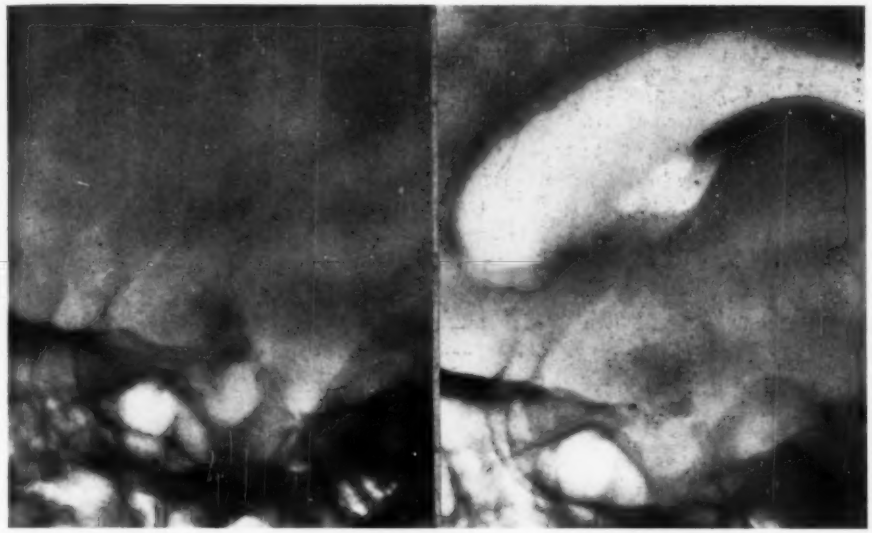


Fig. 14 a. Fig. 14 b.

Fig. 14. Hypothalamus glioma.

a) Preliminary film. »Nodular» calcification. Destruction of the corticalis of the floor of the sella.

b) Ventriculography. Only a small part of the third ventricle near the Monroe's foramen is filled with air. Compare Fig. 9: no differentiation between craniopharyngiomas and suprasellar gliomas is possible in such instances by means of pneumography.

(3.2 per cent). The appearance of the calcification is »nodular» in 1 case and »cystic» in 10 cases. In 8 of these 10 cases the calcification is arranged in such a way that one has the impression that it projects the line of the dorsum sellae which is pushed backwards. Of the other two cases the »cystic» calcification outlines the complete capsule of the tumour in one and a section of the capsule not connected with the sella in the other. The type of the adenoma was chromophobic in all the 11 cases. Lime-salts were actually found in the capsule of the tumours. Encephalography is of no help in differentiating adenomas with calcified capsules from craniopharyngiomas with cystic calcification, and solely by roentgenology, such differentiation is generally impossible. However, we never found increased convolutional markings and separation of the cranial sutures in adenomas.

Suprasellar meningiomas rarely show calcification. CUSHING and EISENHARDT (10) found suprasellar calcification in 2 out of 28 cases of

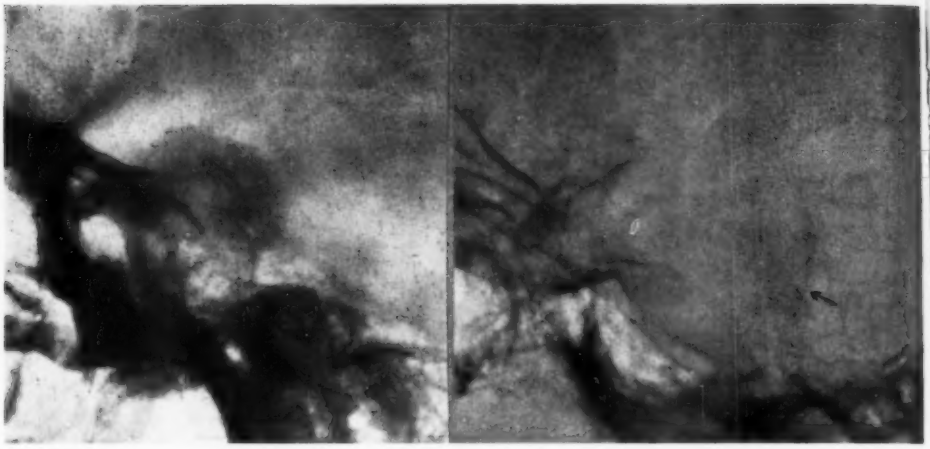


Fig. 15. Suprasellar meningioma. Typical »granular» calcification. There is a 2 cm long hyperostosis immediately in front of the enlarged sella.

Fig. 16. Chromophobe pituitary adenoma. »Cystic» calcification. This projects the line of the dorsum sellae. »Cup-shaped» sella. The capsule of the adenoma was calcified.

suprasellar meningiomas reported in 1938 (7 per cent). In our series of 45 we have found 3 cases with calcification (6 per cent). According to RUPP (33) the calcification of the suprasellar meningiomas should be located more forwards than in craniopharyngiomas. We agree from our experience. The calcified area surmounts the planum sphenoidale. In our 3 cases and in 1 case of DAVIDOFF reported by CUSHING and EISENHARDT (in which it is possible to get information as to the appearance of the calcification), the calcification has the same peculiar appearance. It consists in fact of very small elements compactly and uniformly gathered together so that one has the impression of an homogeneous structure. We call this type of calcification »granular». In our opinion the appearance of this type of calcification prevents diagnostic misinterpretations.

Chordomas have the immediate retrosellar region as their commonest intracranial localization by reason of their origin from the remnants of the chorda dorsalis. EPPLE and RUCKENSTEINER (16) who have studied (1946) the clivus chordomas from the roentgenological point of view state: »Verkalkungen scheinen in Chordomen ausnahmsweise vorzukommen.» CAIRNS and JUPE (5) regarding the intracranial chordoma state that »It is not calcified.» On the other hand in a case reported by ALAJOUANINE, DE MARTEL, OBERLING and GUILLAUME (1) the sella was partly

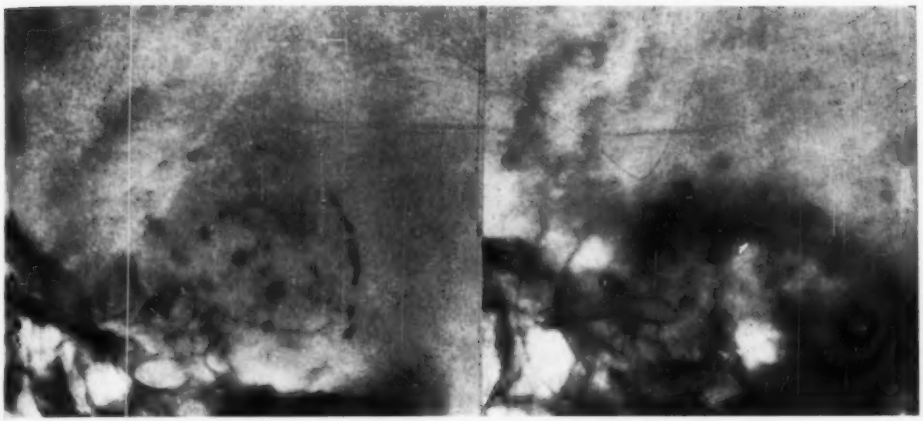


Fig. 17 a. Fig. 17 b.

Fig. 17. Chordomas. Note the »mixed» type of calcification with a dense surrounding line and markedly opaque irregular elements within. In b) the cystic component of the calcification is only recognizable posteriorly. In both cases the sella was practically normal in the lateral views. An half-axial projection revealed however destruction of the left part of the dorsum sellae in both cases.

destroyed and there was suprasellar calcification with highly dense outlines suggesting a bony capsule. We have 4 chordomas in the vicinity of the sella, all calcified. In one case the calcification is strictly suprasellar, while in the other 3 cases the calcification is suprasellar as well as parasellar. The appearance of the calcification is »mixed» in 3 cases and »nodular» in 1. In the 3 cases where the calcification is »mixed», the appearance is peculiar. The line of the cystic component of the calcification is dense and within its limits is included the nodular component formed by elements more irregular and dense than in craniopharyngiomas. Furthermore in these chordomas the intracystic calcifications are present in the entire area delimited by the cystic calcification, and not only in the posterior-inferior part as in craniopharyngiomas with calcification of the »mixed» type. In 3 of the 4 cases the dorsum sellae is destroyed from one side (but seems practically normal in a lateral view of the skull). In 2 cases a part of the clivus is destroyed and in 3 cases the apex of one of the petrous bones. As a rule the appearance and the site of the calcification and of the bone destruction are in our opinion sufficient for a diagnosis of intracranial chordoma.

In our material we have 1 *cholesteatoma* (among 4 cases of verified suprasellar cholesteatomas) showing calcification. This is of the »nodular» type but the single elements are a little larger than in craniopharyngio-

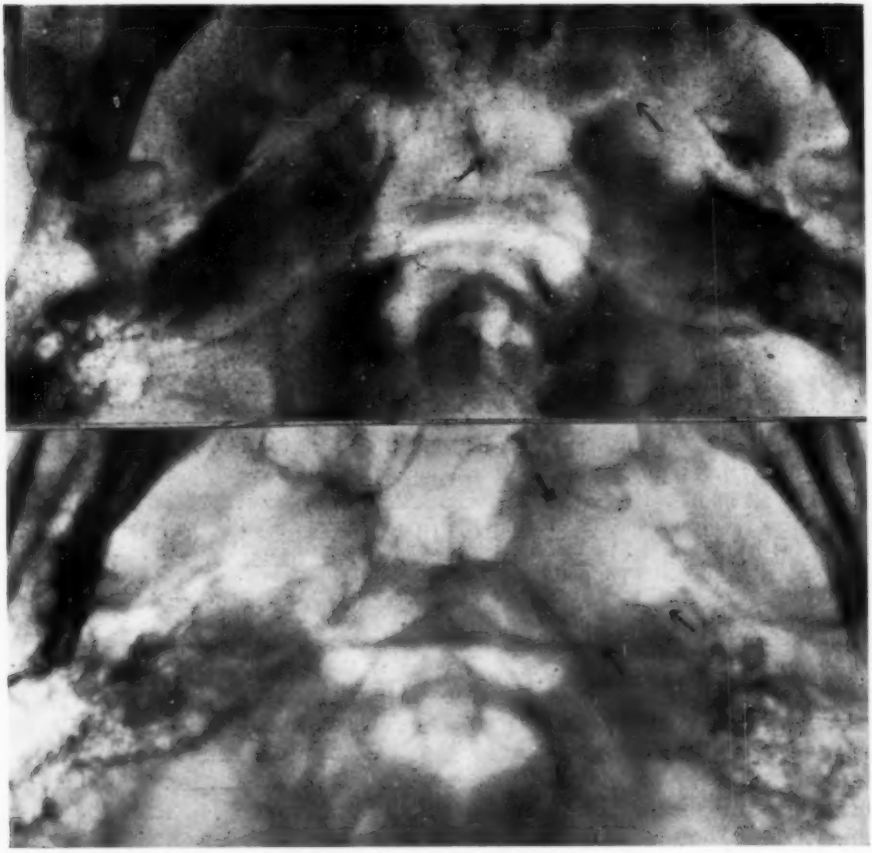


Fig. 18. Chordomas. Axial view. In a) destruction of the clivus on the left together with destruction of the apex of the petrous bone. In b) destruction of the left petrous bone.

mas. OLIVECRONA (30) stated in 1932 that the suprasellar cholesteatomas should be included in the group of suprasellar tumours which do not modify the sella turcica. Nevertheless 2 of our 4 cases show an »increased intracranial pressure» sella. One case has a considerably widened sulcus chiasmatis with enlargement of both optic foramina.

Aneurysms in the sellar region showing calcification are in our experience rare. Of 75 cases we have only 3 with calcification; this is fine, curvilinear and sharply outlined and is always parasellar in site. Thus

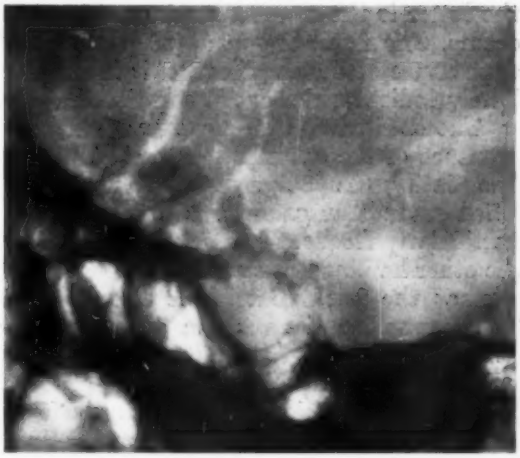


Fig. 19. Cholesteatoma. »Nodular» calcification. The elements of the calcification are however more confluent and dense than in craniopharyngiomas with »nodular» calcification. Sella enlarged with destruction of the corticalis of the anterior aspect of the dorsum.

misinterpretations should be avoidable. Arteriography readily clarifies the nature of the changes.

Occasionally *central gliomas* may extend downwards into the third ventricle and show calcification in their lower part. This calcification thus comes to lie in the suprasellar region and may mislead the radiologist. We have 8 such cases. Nevertheless, contrast examinations, especially pneumography, make the diagnosis obvious.

There is also in our material a case of a subdural haematoma located between the optic nerves and which presents a suprasellar calcification of the »nodular» type. It seems that at present the diagnosis in this and similar cases can only be made at operation.

Conclusions

Suprasellar calcification is by no means pathognomonic of craniopharyngiomas. In addition to craniopharyngiomas — suprasellar gliomas, chordomas, suprasellar meningiomas, pituitary adenomas, cholesteatomas, aneurysms of the internal carotid artery and its branches, and old haematomas — may all show suprasellar calcification.

Two lesions present suprasellar calcification with particular frequency. These are craniopharyngiomas with roentgenologically demonstrable calcification in about 55/60 per cent of cases and suprasellar gliomas (optic chiasma gliomas and hypothalamus gliomas) which are calcified according to our experience in about 50 per cent of cases. The calcification of the craniopharyngiomas is of two types — »cystic» and »nodular» — of which

the latter is the commoner. A combination of the two types sometimes occurs. The fairly frequent (33 per cent of the calcified cases) »cystic» appearance of the calcification is in our opinion the only sign which definitely indicates the cystic constitution of a craniopharyngioma.

The calcification of suprasellar gliomas is »nodular». Neither the appearance of the calcification nor the bony changes give information differentiating craniopharyngiomas with »nodular» calcification from calcified suprasellar gliomas. In our experience the »J-shaped» sella is not particularly frequent in the optic chiasma gliomas. Encephalography on the other hand permits a definite differential diagnosis. In the suprasellar gliomas the third ventricle is altered in shape and pushed backwards. The suprasellar cisterns can generally only be filled with air in their posterior part and if the anterior part be filled, it is pressed against the sellar entrance. An airfilling of the cisterna laminae terminalis is never obtained. In craniopharyngiomas the third ventricle is pushed backwards and upwards and the tumour produces a bulging into the anterior-inferior part of the third ventricle between the chiasmal and infundibular recesses. When the newgrowth attains a large size the change in the appearance of the third ventricle, now nearly completely obliterated, is no longer recognizable. In craniopharyngiomas located within and above the sella, the suprasellar cisterns are lifted and surround the upper part of the tumour. Such a condition never occurs in suprasellar gliomas. In craniopharyngiomas located entirely above the sella the suprasellar cisterns are pushed against the sella but the filling of these cisterns with air is equal in both the anterior and posterior portions, and not as in suprasellar gliomas deficient anteriorly. We did not find combined »cystic» and »nodular» (*i. e.* mixed) calcification in suprasellar gliomas.

In pituitary adenomas with calcified capsules a picture very like that occurring in craniopharyngiomas with »cystic» calcification may occur. Nevertheless the capsule calcification of the adenomas generally projects the line of the dorsum sellae being thus located posteriorly, while the »cystic» calcification of craniopharyngiomas is more likely to be located anteriorly or upwards. A combined supra- and intra-sellar craniopharyngioma with cystic calcification can, with encephalography, give exactly the same picture as calcified adenomas in the same site. Calcification in pituitary adenomas are however rare, occurring in our experience in about 3 per cent of cases.

The differential diagnosis in other lesions which may present suprasellar calcification is easier. Suprasellar meningiomas rarely show calcification. When they do so, it is of a characteristic type and consists of small elements uniformly and very densely arranged together; one has the impression of an homogeneous calcified area located more forwards

than the calcification of craniopharyngiomas and suprasellar gliomas, lying above the planum sphenoidale.

Chordomas are as a rule only partially located in the suprasellar region being mostly parasellar in situation. Their calcification is dense and composed of a linear component (calcified capsule) and of irregular calcified spots inside the capsule. Furthermore chordomas generally destroy the dorsum sellae, the clivus and the apex of the petrous bone.

Suprasellar cholesteatomas showing calcification are not frequent. In these tumours the calcification is very like the »nodular» type craniopharyngiomas although in suprasellar cholesteatomas the calcified elements are a little larger and more irregularly arranged.

The fine curvilinear calcification of the aneurysms of the internal carotid artery and its branches has generally such a typical appearance that no doubt as to the diagnosis should arise. In addition the aneurysmal calcification lies alongside the sella.

Thus the appearance of suprasellar calcification in itself makes the diagnosis clear in a large proportion of cases, though in others encephalography is necessary for a more definite diagnosis. In all cases in which the sellar changes do not clearly point to the location of the tumour (which happens practically only when the sellar changes indicate an intrasellar tumour), encephalography is indispensable for an accurate assessment of the position of the tumour in relationship to the sella and to the brain.

SUMMARY

A series of 54 craniopharyngiomas showed calcification in 30 cases (54 per cent). In 238 cases, gathered from the literature, the figure was 58 per cent. The authors divide the calcifications into three types: nodular, cystic and mixed. Calcification of the nodular type may be found in suprasellar gliomas (optic and hypothalamic). In a series of 14 such cases 7 were calcified. Differential diagnosis is only possible with pneumography. Cystic calcification may seldom appear in pituitary adenomas (3 per cent in this material of 345 cases) and is then in the authors' series always situated as a projection upwards of the dorsum sellae. Other lesions (suprasellar meningiomas, chordomas, cholesteatomas and aneurysms) are easier to differentiate due to the typical appearance of the calcifications and their situation.

ZUSAMMENFASSUNG

Eine Gruppe von 54 Craniopharyngeomen liess Verkalkungen in 30 Fällen (54 %) erkennen. Bei 238 aus der Literatur zusammengestellten Fällen betrug der Anteil 58 %. Die Verfasser teilen diese Art der Verkalkungen in 3 Typen ein: noduläre, cystische und gemischte. Verkalkung vom nodulären Typus kann bei suprasellaren Gliomen (Opticus und Hypothalamus) festgestellt werden. In einer Gruppe von 14 derartiger Fälle zeigten 7 Verkalkungen. Die Differentialdiagnose ist nur durch die Pneumographie möglich. Cystenförmige Verkalkung kann in seltenen Fällen bei Adenom der Hypophyse auftreten (3 % in dem Material von 345 Fällen) und fand sich dementsprechend in der

Beobachtungsgruppe der Verfasser immer als eine Art Vorsprung über dem Dorsum sellae. Andere Veränderungen (suprasellare Meningeome, Chordome, Cholesteatome und Aneurysmen) sind leichter zu unterscheiden infolge des typischen Aussehens der Verkalkungen und ihrer Lage.

RÉSUMÉ

Une série de 54 craniopharyngiomes présentait des calcifications dans 30 cas (54 pour cent). Sur 238 cas relevés dans la littérature le pourcentage était de 58 pour cent. Les auteurs divisent les calcifications en trois catégories, nodulaires, cystiques et mixtes. Les calcifications du type nodulaire se trouvent essentiellement dans les gliomes suprasellaires (optiques et hypothalamiques). Dans une série de 14 cas du genre, 7 étaient calcifiés. Seule la pneumographie permet le diagnostic exact. La calcification cystique serait extrêmement rare dans les adénomes pituitaires (3 pour cent sur ce matériel de 345 cas) et est toujours considérée dans la série des auteurs comme une élévation du dos de la selle. D'autres lésions (méningiomes suprasellaires, chordomes, cholestéatomes et anévrysmes) sont plus faciles à identifier grâce aux radiogrammes typiques de leurs calcifications et à leur localisation.

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FROM THE DEPARTMENT OF ORAL SURGERY (DIRECTOR: PROFESSOR E. HUSTED) OF THE UNIVERSITY HOSPITAL, THE UNIVERSITY INSTITUTE OF PATHOLOGICAL ANATOMY (DIRECTOR: PROFESSOR J. ENGELBRETH-HOLM) AND THE DEPARTMENT OF HISTOLOGY (DIRECTOR: J. J. PINDBORG) OF THE ROYAL DENTAL COLLEGE, COPENHAGEN, DENMARK

FIBROUS DYSPLASIA OR FIBRO-OSTEOMA[1]

Report of a case

by

J. J. Pindborg

In 1937 ALBRIGHT et alii called attention to the relationship between fibrous dysplasia, cutaneous pigmentation, and precocious puberty, and in the following year LICHTENSTEIN named the syndrome polyostotic fibrous dysplasia. Numerous cases of the disease have been published since then. In 1942 it was pointed out by LICHTENSTEIN & JAFFE that a distinction must be made between a polyostotic and monostotic form of fibrous dysplasia. The latter variety might affect any bone of the skeleton, but the manifestation of the lesion in the skull does not appear to have been fully elucidated. In the case to be reported below pronounced sclerosis of certain cranial bones gave rise to differential diagnostic considerations regarding monostotic fibrous dysplasia.

Case Report

A 22-year-old male was referred by his dentist to the Royal Dental College (in 1949) because of a tumour in the right upper jaw. His past history was negative except for ordinary children's diseases. In 1935 (at the age of 8 years) the patient had been admitted to the Otological Department of the Sundby Hospital with a diagnosis of right maxillary osteoma. A microscopic specimen derived from partial excision of the growth showed osteoma spongiosum. A dentist had observed swelling of the right maxilla one year previously.

At the Dental College the patient reported that he had not had any symptoms except for the deformity which in his own opinion had been stationary for a number of years. He exhibited pronounced facial asymmetry with a bony swelling on the right, occupying the area corresponding with the maxilla and the zygomatic arch, and decreasing in the frontal region around the supra-orbital ridge. Fig. 1 shows the marked

[1] Submitted for publication, April 18, 1951.

Fig. 1.

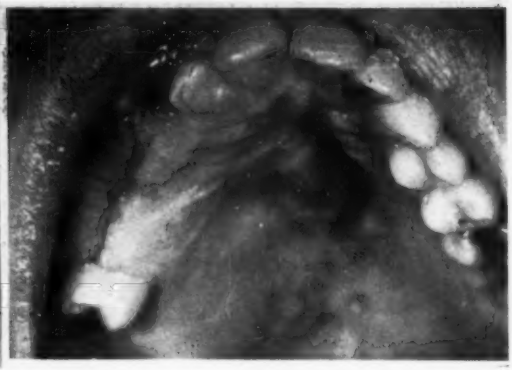

Fig. 2.

Fig. 1. Note the right-sided deformity and the displacement of the upper incisors to the left.

Fig. 2. The oral manifestation of the swelling.

left displacement of the upper incisors. Slight protrusion of the right eyeball, but no diplopia. No abnormality of the skin.

Intraoral examination showed a large swelling involving the entire area of the alveolar process in the right maxilla and occupying a large portion of the palate as well as the alveolabial sulcus (Fig. 2). The tumour was hard to the touch but the mucosa normal. Marked left displacement of the raphe palati. Only 5 teeth were absent, viz. 6, 4, 3 + and — 5[1]; the two first-mentioned were removed because of chronic apical paradentitis. No obstruction to the passage of air through the right nostril.

Roentgen examination of the skull showed very marked changes: »In the lateral view (Fig. 3) there is enormous thickening of the entire base of the skull, extending from the lower part of the frontal bone backwards and occupying the lower portion of the occipital bone almost as far as the lambdoid suture. The base of the skull is of an abnormal thickness, thus the occipital bone is up to 4 cm. thick and the floor of the anterior cranial fossa 3—3½ cm. The bone structure is very dense and homogeneous without spongy trabeculations and appears to be made up of a firm, solid mass. Normal sella turcica and normal pneumatization of the inner ear. There is presumably question of external as well as internal thickening of the bone. In the antero-posterior view (Fig. 4) the thickening is seen to be typically unilateral, only involving the right side on which the dense bone structure extends down to the facial bones, invading the nose as well as the maxilla. The right orbit is perceptibly smaller than the left and its contours are ill-defined; it measures only 3½ cm. in height compared with 4 cm. on the opposite side. Medially, the bony condensation reaches the nasal bone and the fronto-sphenoidal process, involving the conchae, causing marked enlargement, particularly of the superior one, and displacing the nasal septum far to the left. Moreover, the dense bone structure occupies the entire right maxillary antrum which lacks pneumatization; this is apparent

[1] According to the HADERUP system of dental designation, + signifies the upper teeth and — the lower teeth. If the symbol be placed to the right of the figure, the right side is indicated, and vice versa. 0 before the figure indicates a deciduous tooth.

Fig. 3. Lateral film of the skull. Note the enormous thickening of the entire base and the condensation in the upper jaw.

also on the lateral view. The bony condensation also affects the hard palate and the right alveolar process. The growth is expansive — the maxilla projecting far to the right — and extends downwards and, as mentioned above, upwards to the orbit, and medial to the nose. At the same time there is a marked forward projection of the maxilla. On the left, there are no definite changes in the maxilla or of the theca cranii itself. Further roentgen examination did not reveal any abnormality of the vertebral column, bony thorax, pelvis, or extremities.

In other words, this is a case of gross cranial changes, the base of the skull and the right facial bones, except for the mandible, being the site of pronounced, infiltrative and expansive thickening of the bones of an extremely dense and homogeneous structure. Judging by the roentgen films, it seems to be a widespread osteoma of diffuse growth. The site and the expansive growth correspond exactly with the findings in this form of growth, although the extent is of quite exceptional dimensions. A fibro-osteoma is not out of the question, but the two conditions cannot be distinguished roentgenographically. The appearances do not suggest malignancy, since there are no destructive changes. Fibrous dysplasia would probably not involve such marked sclerotic changes and Paget's disease usually affects the theca cranii and shows a looser bone structure than in this case» (sd. GREGERS THOMSEN). A comparison between antero-posterior films obtained in 1938, 1939 and 1949 showed increased sclerosis in the right frontal bone, whereas the other bony changes appeared to be unaltered.

In order to provide space for a partial denture to replace the absent upper teeth, a partial resection of the maxilla was performed in the Department of Oral Surgery of the University Hospital. The removed tissue was examined microscopically and showed »spongy bone tissue (Fig. 5) the medullary substance of which has been replaced by connective tissue. The latter is in places acellular (Fig. 6) showing an abundance of fibrils, but in other places it is more cellular with a tendency to whorl formation

(Fig. 7). Nevertheless, the cells are uniform and show no signs of malignant proliferation. The tissue contains a good deal of large, thin-walled blood-vessels. No signs of past haemorrhage or cyst formation. In several places, the bone tissue exhibits some rarefaction, but no other definite abnormalities. Extremely few osteoclasts and no exudative inflammatory changes. Should this be an osteoma, it would have to be one with violent, secondary connective-tissue proliferation, and this does not appear likely. It is probably not a case of true tumour formation especially as there are no signs of malignant proliferation. The appearances are compatible with fibrous dysplasia of bone. The absence of osteoclastic activity contradicts hyperparathyroidism. Histologically, Paget's disease cannot be ruled out with certainty, but this diagnosis is unlikely in view of the very atypical findings, the site of the lesion, and the patient's age. Therefore, the histological diagnosis must be: spongy bone tissue in which the bone marrow has been replaced by connective tissue (presumably fibrous dysplasia of bone)» (sd. W. KLÆR).

Fig. 4. Antero-posterior view. Note the typically unilateral thickening of the bones.

Discussion

Thus, the condition was interpreted as being an osteoma (fibro-osteoma) by the radiologist and as fibrous dysplasia by the pathologist. Before choosing between the two alternatives, the writer will briefly review some morbid conditions which may give rise to the signs in question.

The unilateral deformity was distinctly visible upon inspection, a fact which brings *leontiasis ossea* to mind. It is still being discussed, whether this is interpretable as an independent, well-defined entity (*e. g.* TRAUNER (1942) and THOMA (1944)) or whether it is merely a collective term for sclerotic osseous changes in various diseases (int. al. FREEDMAN (1933) and WEINMANN & SICHER (1947)). BOENINGHAUS (quoted by LIECHTI (1946)) applies the term leontiasis ossea to sclerotic processes confined mainly to the bones of the face and the base of the skull and gives the diagnosis of *diffuse osteoma* in parentheses. In our differential diagnostic argument we have now reached the tumours. Osteoma, in the ordinary sense of the word, may be ruled out in this case, as the process is spread diffusely in the entire base of the skull and its histological architecture is not compatible with an osteoma. BOCKENHEIMER (1908) used the expression diffuse hyperostosis, and

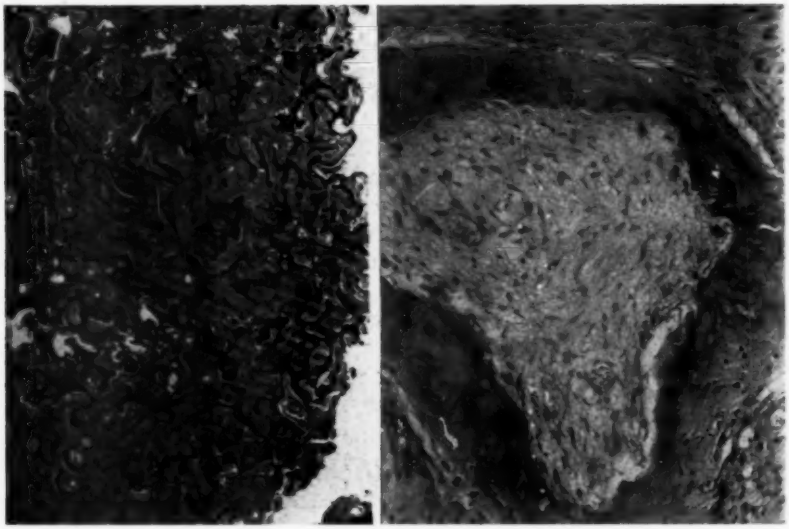

Fig. 5. Fig. 6.

Fig. 5. The removed tissue. Note the spongy bone structure.
Fig. 6. Higher magnification of a relatively acellular area of the removed tissue. Original magnification: 10 × 12.

GILHEPSY (1927) called the same lesion »chronic hyperplasia of the upper jaw».

In 1946 BILLING & RINGERTZ reported 25 cases of fibro-osteoma, defined as a tumour »having a ground substance of connective tissue, in which clumps and spicules of bone tissue are formed». They made a distinction between the more common type of localized fibro-osteoma and the rare variety of diffuse spread (called leontiasis ossea in the literature). Although the authors employ the term fibro-osteoma, they realize that it is doubtful whether »these formations are true tumours», stating that they are more readily interpretable as »a growing malformation». In the present writer's opinion it is doubtful whether the term tumour is correct for such diffuse processes. BILLING & RINGERTZ's Cases 22—24 leave no doubt that these authors would have interpreted the present case as one of diffuse fibro-osteoma. Under the heading of »differential diagnosis» they do not even mention the possibility of fibrous dysplasia, although their histological findings were virtually identical with what was described as being typical of fibrous dysplasia. On the other hand, they make a sharp distinction between ossifying fibroma and fibro-osteoma, the former being without direct connection with the skeleton. The relation of ossifying fibroma to fibrous dysplasia

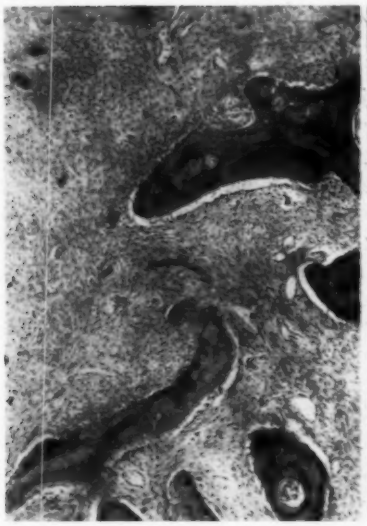

Fig. 7.

Fig. 8.

Fig. 7. Whorl-formation in the connective tissue. Original magnification 10 × 4.
Fig. 8. Metaplastic transformation of the connective tissue into bone tissue. Original magnification 10 × 12.

has been discussed by int. al. SCHLUMBERGER (1946) and MAMMEL (1948) both of whom consider ossifying fibroma to be a variant of fibrous dysplasia.

The cranial changes in *polyostotic fibrous dysplasia* were pointed out as early as 1937 by ALBRIGHT et alii and in 1938 by LICHTENSTEIN and have later been discussed in detail by other workers. Thus, FALCONER & COPE, in 1942, reported two cases of polyostotic fibrous dysplasia exhibiting unilateral bony condensation of the frontal, sphenoidal, ethmoidal, occipital, and maxillary bones. FALCONER & COPE, DOCKERTY et alii (1945), and PUGH (1945) have emphasized that the manifestation of fibrous dysplasia in some parts of the skull differs from the lesions in other bones. In the theca cranii, occipital bone, and mandible, the roentgen appearances are like those in other bones, *i. e.* expanded bone and sclerosis of some areas but with translucencies as the predominant feature. In the frontal, sphenoidal, ethmoidal, and maxillary bones, on the other hand, there are marked, sclerotic processes. In WINDHOLZ' (1947) opinion the peculiar sclerotic cranial changes are due to the dynamic actions to which the skull is exposed. Other workers, *e. g.* SUMMERFELDT & BROWN (1939), FINKLER & COHN (1947), and SANTÉ et alii (1948) have reported polyostotic fibrous dysplasia with cranial changes

Fig. 9. Lamellar bone tissue in the peripheral area of the removed tissue. Original magnification 10 × 4.

almost identical with those encountered in the present case.

LICHTENSTEIN & JAFFE in 1942 called attention to the fact that the *monostotic variety of fibrous dysplasia* had previously been called localized osteitis fibrosa. In their opinion the lesion is not as rare as previously believed. Among 23 cases of their own, they found 9 to affect single bones. The figure found by SCHLUMBERGER (1946) was 67 out of 69 cases. Among his 67 monostotic cases 21 % were localized in the calvarium, maxilla, or mandible. WINDHOLZ (1947) observed cranial changes in 10 % of his monostotic cases. Actually, it is not always correct to employ the term monostotic for cranial manifestations, since several bones (frontal, ethmoidal, sphenoidal, and occipital bones) are often involved. The reason why it is used is no doubt that due to their situation the various cranial bones are virtually »fused» to form one bone. In addition to WINDHOLZ (1947), LIECHTI (1946) and JACOBSEN & VRAA-JENSEN (1949) have dealt with these cranial changes. In the last-mentioned authors' Case 5 the lesion involved the frontal, ethmoidal, parietal, and occipital bones as well as the base of the skull, very much like as in the present case. Owing to the facial deformity often attending both the polyostotic and monostotic variety, certain workers (PUGH (1945), LIECHTI (1946), and WINDHOLZ (1947)) consider leontiasis ossea to be interpretable as the cranial manifestation of fibrous dysplasia.

The histopathology of monostotic fibrous dysplasia does not appear to differ essentially from that of the polyostotic variety. The primary changes consist of widespread conversion of the bone marrow into connective tissue which may contain many cells, arranged in entwined bundles and whorls (Fig. 7) or connective tissue with few cells (Fig. 6) containing an ample content of collagen. In the connective tissue there is new bone formation, apparently by metaplasia of the already existing connective tissue (Fig. 8). A homogeneous substance is deposited between the fibrils and gradually enclosed in the cells after which calcium is deposited in the new-formed bony matrix. At a later stage there may be organization of the primitive bone tissue, giving the bone a lamellar structure (Fig. 9).

It will be seen from what has been stated above that changes like those observed in the present case have previously been called leontiasis ossea, diffuse osteoma, fibro-osteoma, or fibrous dysplasia. In the case under discussion, the diagnosis of leontiasis ossea does not appear applicable, partly because of the unilateral character of the lesion, and partly because of the ample connective tissue demonstrated in the histological specimens. Applying a term suggesting a tumour, such as diffuse osteoma or fibro-osteoma, to processes so diffuse as those in question would be vitiating our present conception of a tumour. This leaves the diagnosis of fibrous dysplasia; since the lesion was confined to the skull, the polyostotic variety can be ruled out (with the reservations previously mentioned). Instead of calling this special manifestation monostotic fibrous dysplasia the writer suggests the term *diffuse fibrous dysplasia of the cranial bones* by the use of which it could be distinguished from monostotic lesions in other situations.

SUMMARY

Gross sclerotic processes in the maxillary, frontal, and occipital bones and the entire base of the skull were observed in a male, aged 22. The changes did not involve other bones, and there were no signs of generalized disturbances. Histological examination showed mainly cellular connective tissue with metaplastic new bone formation. The different terms used in the course of time for similar lesions are reviewed with particular reference to fibro-osteoma and fibrous dysplasia. No doubt, the condition is interpretable as fibrous dysplasia. Owing to its situation the writer suggests the term: diffuse fibrous dysplasia of the cranial bones.

ZUSAMMENFASSUNG

Bei einem 22 Jahre alten Mann wurden ausgedehnte sklerotische Prozesse in den Knochen des Oberkiefers, der Stirn, des Hinterhauptes und der gesamten Schädelbasis beobachtet. Andere Knochen wurden von der Veränderung nicht ergriffen. Es bestanden keine Zeichen für eine allgemeine Störung. Die histologische Untersuchung liess in der Hauptsache zelluläres Bindegewebe mit metaplastischer Knochen-Neubildung erkennen. Die verschiedenen Krankheitsbezeichnungen, die im Laufe der Zeit für ähnliche Veränderungen gebräuchlich waren, werden besprochen unter besonderer Berücksichtigung des Fibroosteoms und der Dysplasia fibrosa. Ohne Zweifel kann der beschriebene Fall als Dysplasia fibrosa erklärt werden. Wegen der örtlichen Anordnung der Veränderungen schlägt der Verfasser die Bezeichnung vor: Dysplasia fibrosa diffusa der Schädelknochen.

RÉSUMÉ

Un processus sclérotique de grande envergure a été observé chez un homme de 22 ans. Ce processus s'étendait aux os maxillaires, frontaux et occipitaux et à la base entière du crâne. Les altérations n'intéressaient pas d'autres régions et il n'y avait aucun signe de troubles généralisés. L'examen histologique montra principalement un tissu connectif cellulaire avec néoproduction osseuse métaplastique. Les appellations variées employées au cours des années pour des lésions similaires sont passées en revue; spécialement en ce qui concerne les fibrostéomes et la dysplasie fibreuse. Arguant de la localisation l'auteur propose le terme de dysplasie fibreuse diffuse des os du crâne.

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FROM THE SURGICAL DEPARTMENT (DIRECTOR: PROFESSOR J. HELLSTRÖM) AND THE ROENTGEN DEPARTMENT (DIRECTOR: PROFESSOR Å. ÅKERLUND) OF KAROLINSKA SJUKHUSET, STOCKHOLM, SWEDEN

URETHROCYSTOGRAPHIC STUDIES OF NERVOUS DISTURBANCES OF THE URINARY BLADDER AND THE URETHRA[1]

A preliminary report

by

G. Giertz and *K. Lindblom*

For many years experimental and cystoscopical studies of the micturition mechanism have been carried out. The roentgenographic completion is somewhat new.

BARRINGTON, in 1914, was able to demonstrate that the division of the sacral posterior roots in cats caused retention of urine with increased laxity of the bladder and dilatation of the urethra as far up as to the prostate, the »funnel urethra», while the remaining part of the posterior urethra was tightly closed. In addition, there was a continuous dilatation of the rectum which in one case led to death from faecal impaction.

ALEXIEFF, in 1914, and SCHRAMM, in 1920, by cystoscopy, found the bladder neck constantly open, the former in cases of enuresis nocturna, the latter in lesions of the lower part of the spinal cord, in tabes, and in disseminated sclerosis. SCHRAMM described the open bladder neck combined with trabeculation and diverticula in the bladder in his cases; the ureters and kidney pelves were often dilated. SCHRAMM in 1926 added that the urethral widening extended downwards to the external sphincter.

SCHWARZ, in 1922, considered the funnel urethra to be caused by an external sphincter spasm. GOLDENBERG, in 1924, added that the funnel urethra indicated a spinal lesion only when combined with the other changes described by SCHRAMM, such as a trabeculated bladder; a functional irritation might also cause the urethra to remain open.

[1] Read before the Staff of the Mayo Clinic, Sept. 19, 1950; submitted for publication March 12, 1951.

EPSTEIN and JUSCHELEVSKY and MICHELSON in 1929 published a list of cases with the Alexieff-Schramm sign. In those cases in which the site of the nerve lesion was known, this sign accompanied the spinal or root lesions.

In 1945 JACOBSON described experimental dysfunction of the bladder in dogs. Division of the posterior sacral roots or ganglia resulted in an *atonic bladder* with low tonus, large capacity and absence of contractions. Lesion of the sacral portion of the cord, the cauda, or the pelvic nerves produced an *autonomous bladder* with high tonus, low capacity, accompanied by small 'autonomous jerks' in the cystometrogram; and later a muscular hypertrophy »as a consequence of the autonomous activity of the detrusor muscle». Lesions of the cord above the sacral level produced an *automatic, or reflex, bladder* with increased tonus, lowered capacity, and large »automatic» contractions in the cystometrograms; voluntary control and vesical sensation were absent.

PRATHER and PETROFF in 1947 published a urethrocystographic study of the bladder neck in 129 cases of spinal cord injuries. They also found a dilatation of the bladder neck in cases of cervical injuries. The condition at the external sphincter and the occurrence of trabeculation was not mentioned.

EMMETT and DAUT and DUNN in 1948 demonstrated the »automatic» bladder with external sphincter spasm relaxing after pudendal block.

The most extensive description of the urethrocystographic appearance of the neurogenic bladder was recently published by NEY and DUFF. They described: the funnel urethra, the pine-tree shaped bladder, the vesico-ureteral reflux, the hour-glass bladder, saccular dilatation of the posterior urethra, pseudosphincter formation below the bladder neck, the spastic external sphincter, the relaxed external sphincter, the contracted hypertonic bladder and the large hypotonic bladder. They leave the reader in doubt as to the site of the nerve lesions which produced the various signs.

Purpose, Material and Technique

The purpose of this study is to differentiate urethrocystographic appearances which can be referred to definite sites of nervous injuries or diseases.

The collection of material began in 1940 when micturition films were introduced as a supplement to ordinary retrograde urethrocystography. In spite of the long time required to collect the material, it is in many respects still too small. This report is therefore necessarily a preliminary one.

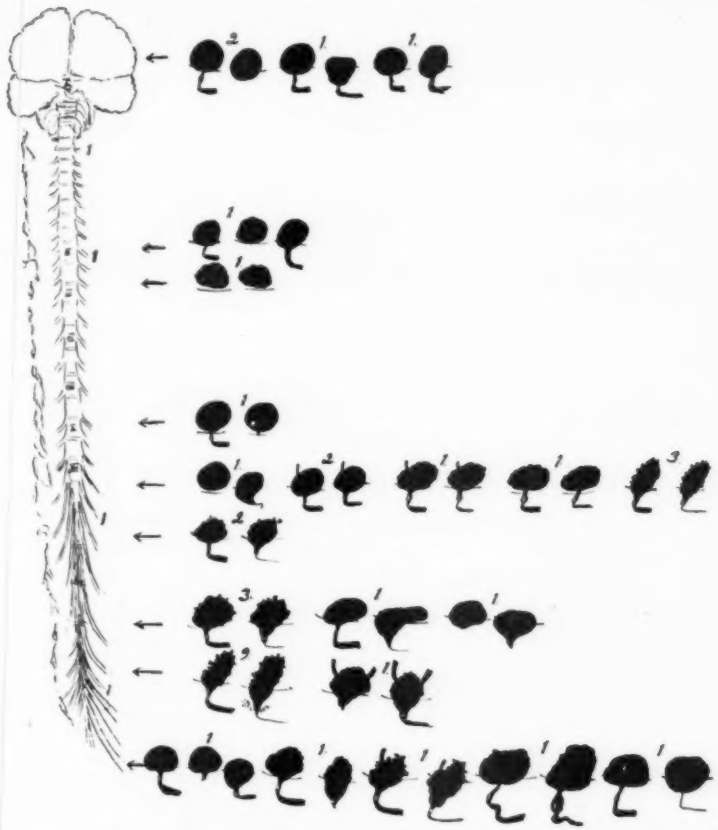

Fig. 1. Site of the nervous lesion and appearance of the bladder and urethra in 37 cases. Figures above the sketches indicate the number of cases with the same appearance.

The investigation of the material was carried out by an urologist and a roentgenologist. The team will continue the work in collaboration with a physiologist and a neuro-histologist.

The technique was described by EDLING in 1944. It was necessary to obtain films even if no urine or a few drops only were passed on micturition. In many cases a large quantity of dye was needed for micturition. As, in most instances, the voiding pictures had to be taken in the erect posture air was found to amplify the distension of the bladder and thus conserve dye. Stimulation of the detrusor muscle by drugs was valuable in many cases. Finally, patience was absolutely necessary.

Seventy nine cases of nervous disturbances of the bladder and the urethra were satisfactorily examined in this way. In 37 of these the level of the nervous lesion was known. The cases were grouped in accordance with the site of the lesions (Fig. 1). This grouping was essential for the differentiation of the urethrocystographic types in nervous lesions.

Urethrocystographic Types in Nervous Lesions

In this material four main types of urethrocystographic appearences in nervous lesions may be differentiated. These types are illustrated in Figs. 2—7.

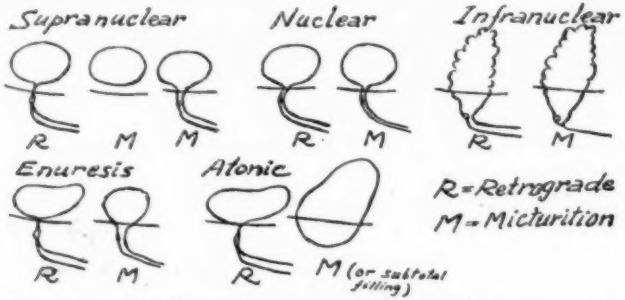

Fig. 2. Urethrocystographic types of nervous disturbances of bladder and urethra.

A. *The supranuclear type* (Fig. 3) (includes cerebral lesions and spinal lesions above the bladder centra) is characterized by a permanently round bladder of moderate capacity with a decreased tendency to open the bladder neck, and by an unchanged shape during voiding. The bladder is often moderately trabeculated. If the patient be able to strain, the level of the bladder may be lowered to some extent. If the patient be able to micturate, and this act may sometimes be accomplished by irritating the skin surface in the bladder region, the urethra opens. The opening may be wider than normal without spasm of the external sphincter. The emptying of the bladder is usually incomplete, leaving residual urine.

B. *The infranuclear type* (Fig. 4) (includes spinal lesions below the bladder centra, mainly lesions of the cauda equina) presents a »pine-tree shaped bladder» with a »funnel urethra» terminating with a marked narrowing of the lumen at the level of the external sphincter. The narrowing usually increases during voiding until there is total obstruction producing a threadlike lumen. The bladder is permanently raised and in most cases markedly trabeculated with small diverticula. Frequently the ureteral orifices are open causing backflow and ureteral and pelvic dilatation.

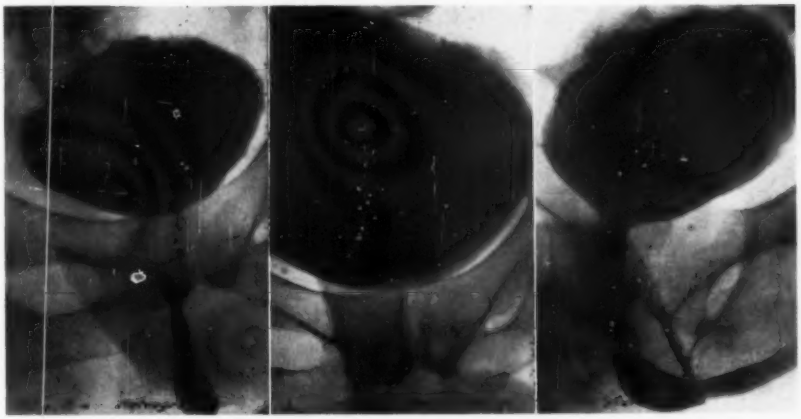

Fig. 3 a—c. Urethrocystograms in transverse lesion at level of Th 1—2. Supranuclear type.

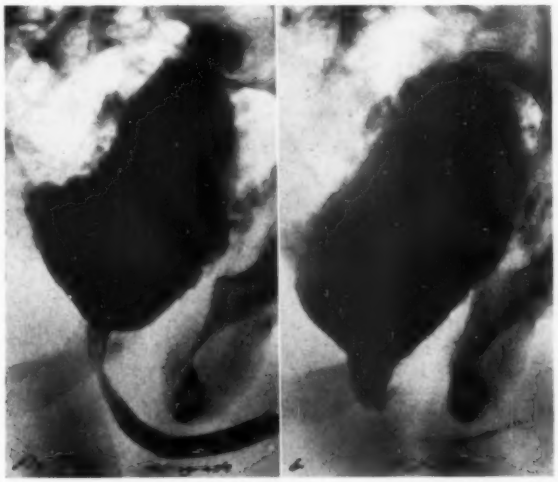

Fig. 4 a—b. Urethrocystograms in lumbosacral meningocele. Infranuclear type.

At the level of the bladder centra, corresponding to the Th 12 and L 1 vertebrae, the infranuclear type occurred in 5 out of 7 cases. The remaining two cases differed from the pure supra- and infranuclear types: the shape of the bladder was similar to that in the supranuclear type but the bladder neck was constantly open as in the infranuclear type

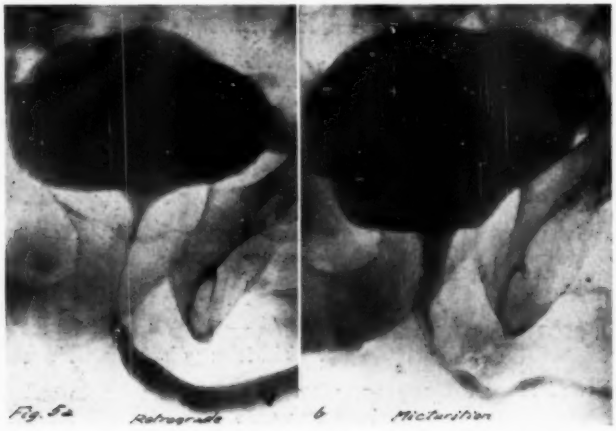

Fig. 5 a—b. Urethrocystograms in transverse lesion at level of Th 12—L 1. Nuclear type (See text).

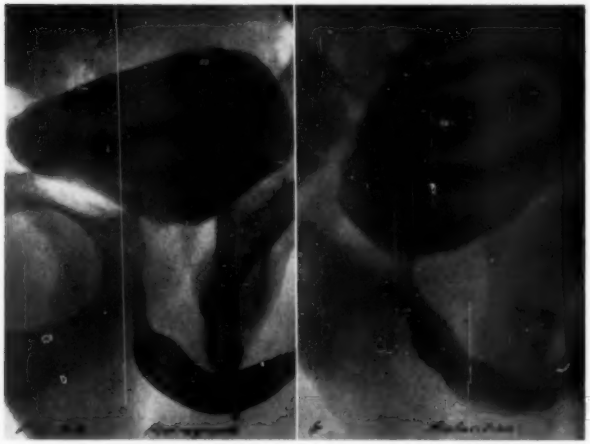

Fig. 6 a—b. Urethrocystograms in enuresis nocturna since birth. Enuresis type.

though not widened more than during normal micturition. There were no signs of external sphincter spasm. These two cases might represent a separate *nuclear type* (Fig. 5).

C. *The enuresis type* (Fig. 6) appears normal during the retrograde filling but has an increased tendency to empty; during micturition the

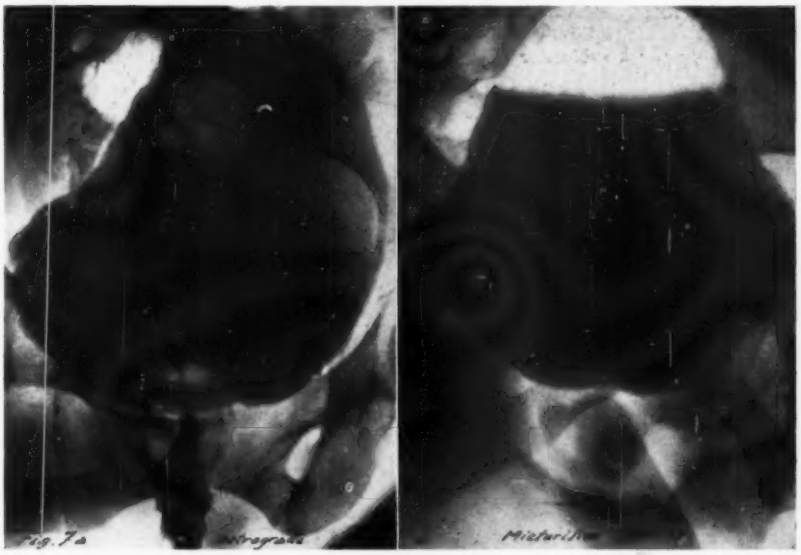

Fig. 7 a—b. Urethrocystograms in female. Atonic type; bladder capacity, 1,000 cc without increased intravesical pressure.

urethra widens more than normally. Rarely, the bladder shows slight trabeculation and single diverticula are present.

D. *The atonic type* (Fig. 7) characterized by a large hypotonic bladder also appears normal during the retrograde filling but has little tendency to empty. In most cases it is impossible to obtain a micturition picture. The bladder has to be greatly distended with dye before the urethra opens. Micturition is facilitated by the erect posture. In that position, dye may be supplemented by air filling of the upper part of the bladder. During voiding the urethra does not open more than normally. There is no spasm of the external sphincter.

Urethrocystographic Types in Relation to the Site of the Nervous Lesion.

The reader is referred to the table in Fig. 8. A study of this table shows beyond doubt that there is a distinct difference in the urethrocystograms in lesions above the Th 12 vertebra and below the L 1 vertebra. These two sites are characterized by the supranuclear and the infranuclear type respectively.

In addition, the table demonstrates that, as a rule, the atonic type is not caused by a lesion of the central nervous system. This agrees with

Known site of lesion	Number	Types					
		Supra-nuclear	Nuclear	Infra-nuclear	Enuresis	Atonic	Undef-inite
Above Th 12	7	7 (2E)	—	—	—	—	—
Level Th 12—L 1	8	—	2	5	—	—	1
Below L 1	17	—	—	16	—	—	1
Pelvis	5	—	—	2	—	—	3
Unknown site of lesion							
Lues	9	4 (1E)	—	5	—	—	—
Poliomyelitis	4	2 (2E)	—	2	—	—	—
Other cord lesions	6	4 (2E)	—	—	—	2	—
Enuresis	11	—	—	—	10 (10 E)	—	1
Retention	14	—	—	—	—	12	2

E = Enuresis type during micturition.

Fig. 8. Number of cases of different urethrocystographic types in relation to site of nerve lesion.

our clinical experiences in which megacystis (atonic bladder) was most often a complication of pregnancy, of delivery, or of abdominal operations and especially those on the minor pelvis.

Another point of interest in the table is the appearance of the enuresis type during micturition in cases of nervous lesions above the 12th thoracic vertebra. This fact may indicate that true enuresis nocturna is caused by a lack of cerebral control of the spinal cord centra of the bladder. However, there is a urethrocystographic difference between the cerebrospinal injury and true enuresis nocturna; the latter disease presents a normal picture in the retrograde filling and there is good ability to initiate micturition and to complete emptying of the bladder.

Cases of cystitis and vesiculitis may appear similar to the enuresis cases, indicating that a local irritation may play an additional rôle in true enuresis.

The group of indefinite types is still too small to be discussed. There are cases with a wide open posterior urethra down to the colliculus or with a tendency to contract at this level.

The External Sphincter Spasm. Its Rôle in Trabeculation. Comparative Anography. Blockage of the External Sphincter Spasm.

A separate study was made of external sphincter spasm in cases of spinal lesions below the first lumbar vertebra, the infranuclear type.

During the retrograde injection there was no more resistance than normal and the lumen in the region of the external sphincter opened in the usual manner. However, during micturition it closed almost completely by reason of an external sphincter spasm while the lumen above

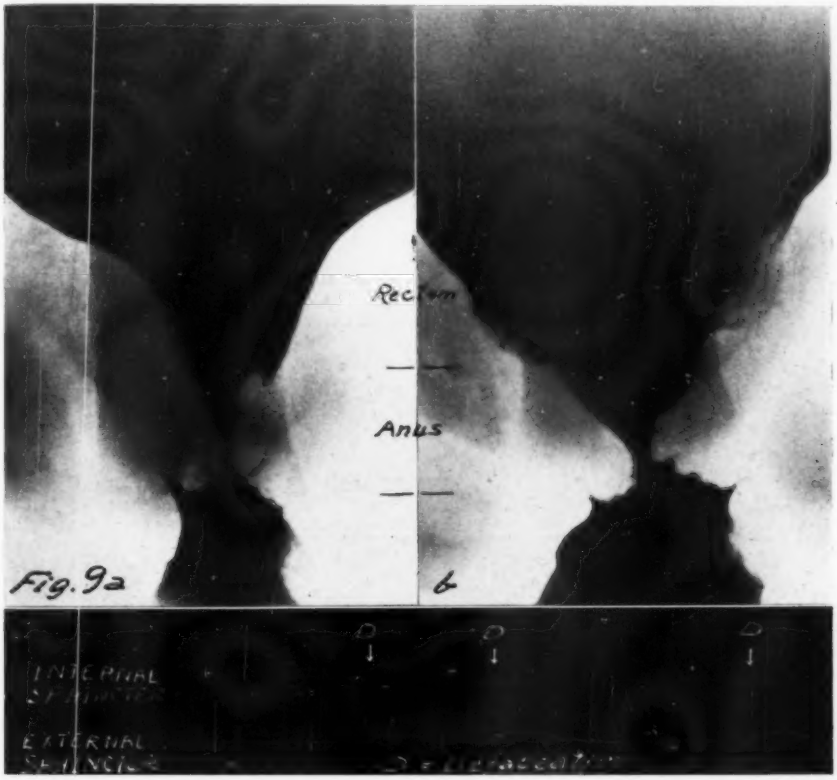

Fig. 9 a—b. Anograms in a case with infranuclear type of nerve disturbances of urinary bladder after disc operation at L 4 level. Below: records of sphincter tonus in same case.

this level was almost constantly wide open. If it be true that the presence of urine in the upper part of the urethra irritates the detrusor muscle the constantly open bladder neck may be partly responsible for the raised form of the bladder. The trabeculation is explained by the obstruction to the outflow by the external sphincter spasm. If the trabeculation be only slight it may be due to a backflow through open ureteral orifices, which decreases the bladder pressure.

In cases of a micturition spasm of the external urethral sphincter there was also a defaecation spasm of the external anal sphincter (Fig. 9). The defaecation spasm was demonstrated by anography, which means roentgenography of the anus in the resting state and during defaecation,

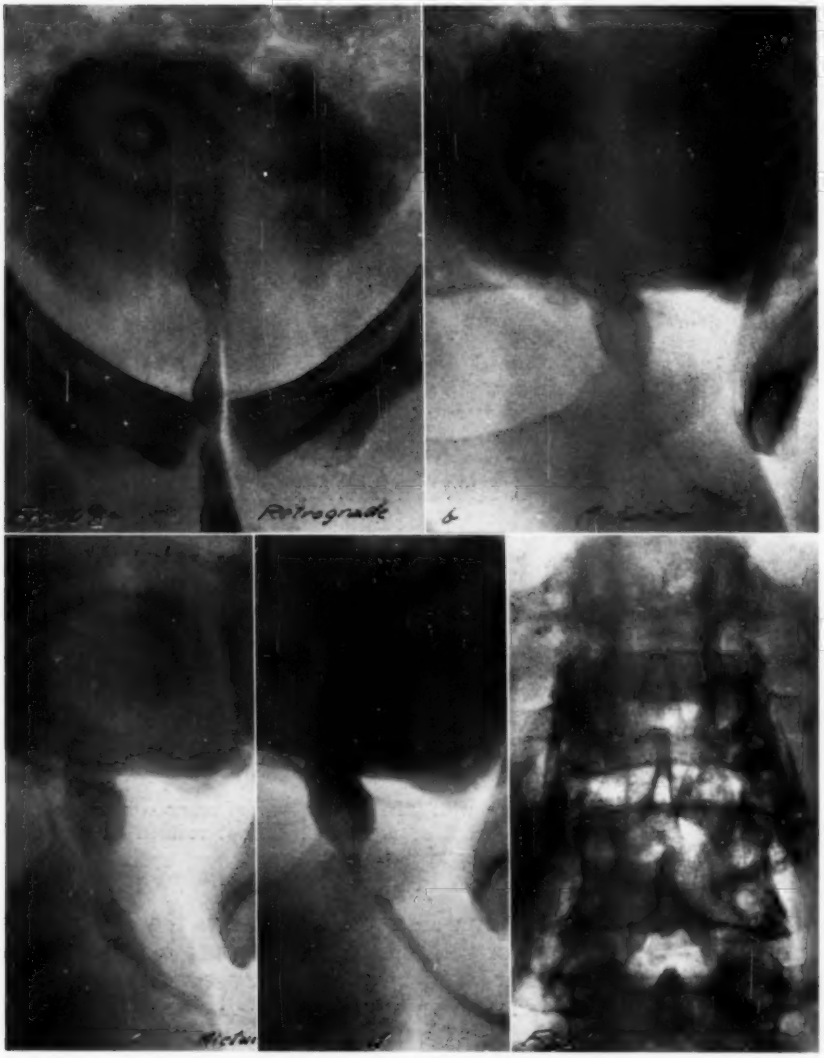

Fig. 10. a—d. Urethrocystograms in a case with infranuclear type of nerve disturbances after poliomyelitis inferior. In Fig. 10 c relaxed sphincter after etamonin. In Fig. 10 d partial relaxation of external sphincter after paravertebral anaesthesia in the upper lumbar region.

Fig. 11. Roentgenogram demonstrating site of paravertebral anaesthesia in same case as in Fig. 10. The novocain was mixed with perabrodil.

after the rectum has been filled with a barium paste. The anal spasm was easily felt by the finger during defaecation. It was also recorded by two small balloons in the anus, one in the inner and one in the outer part of the canal.

Finally, roentgenographic studies were made of a blocking of the spasm of the external urethral sphincter. The spasm ceased after intramuscular injection of etamonin (Fig. 10 c). It was also demonstrated that paravertebral injections of novocain in the lumbar region (Fig. 11) definitely caused the spasm to decrease (Fig. 10 d), indicating that the reflex for sphincter closure passes through the paravertebral part of the autonomous nervous system.

During the blockage of the external sphincter spasm the cases behaved similar to the two cases of the presumed nuclear type: the patients micturated simply and easily by increasing the intra-abdominal pressure. This is interesting, not only as a suggestion of a way in which the deleterious spasm might be treated, but also because it indicates that the spastic reflex may have to pass through or originate from the region of the bladder centra of the spinal cord. It seems to be the external sphincter spasm and its sequelae — trabeculated bladder and markedly widened bladder neck — which form the roentgenographic difference between the infranuclear type and the presumed nuclear one, *e. g.* the lesions below and in the bladder centra of the spinal cord.

Conclusions

Urethrocystography during retrograde injection and micturition is a valuable help in the study of nervous disturbances of the bladder and the urethra.

Lesions above and below the spinal cord centra of the bladder have different characteristics.

Lesions above the cord centra have some similarity to true enuresis nocturna indicating that this disease, at least partly, is derivated from a lack of cerebral control of the bladder cord centra.

The atonic bladder is, as a rule, not caused by a cerebro-spinal or nerve-root lesion.

Micturition spasm of the external urethral sphincter is responsible for the obstruction to the outflow and for the marked trabeculation present in root lesions below the spinal cord centra of the bladder. There is an equivalent spasm of the external anal sphincter, demonstrable by anography.

Micturition spasm may be blocked by etamonin or novocain anaesthesia of lumbar paravertebral nerves and ganglia, indicating a method of therapy.

SUMMARY

Urethrocystographic studies were made in seventy-nine cases of nervous disturbances of the urinary bladder and the urethra. It was demonstrated that the level of the nervous lesion had a definite influence on the type of the urethrocystographic changes.

ZUSAMMENFASSUNG

Urethrocystographie in 79 Fällen von nervösen Störungen der Harnblase und der Urethra zeigten, dass das Niveau der nervösen Läsion eine bestimmte Einwirkung auf den Typus der urethrocystographischen Veränderungen hat.

RÉSUMÉ

L'urétrocystographie dans 79 cas de troubles vésicaux et uréthraux d'origine nerveuse a démontré que la site de la lésion nerveuse a une influence prononcée sur le type des changements urétrocystographiques.

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FROM THE ROENTGEN DEPARTMENT (DIRECTOR: DR. CARL SANDSTRÖM),
ST. ERIK HOSPITAL, STOCKHOLM, SWEDEN

CALCIFICATIONS OF THE INTERVERTEBRAL DISCS AND THE RELATIONSHIP BETWEEN VARIOUS TYPES OF CALCIFICATIONS IN THE SOFT TISSUES OF THE BODY[1]

by

Carl Sandström

Calcifications of the intervertebral discs were mentioned for the first time by LUSCHKA in his textbook of anatomy. They were demonstrated roentgenologically in cadavers by BENEKE in 1897, and were first described in human beings in 1922, by CALVÉ and GALLAND. SCHMORL was the first to make thorough pathological-anatomical investigations of these calcifications, which have since been discussed in numerous works by clinicians and roentgenologists. The etiology, and the manner in which they arise, is unknown. In the literature, according to SCHMORL, the etiology is presumed to be traumatic (BARSONY and POLGAR, LYON and others), or to involve persistent overloading (HORENSTEIN and others (cit. SCHMORL)), inflammation (LYON and others), senile alterations (NORLÉN, among others), or some disturbance of the calcium metabolism. SCHMORL himself believes that they arise secondarily to small ruptures with areas of necrosis which become calcified in the intervertebral discs and that in addition some general disturbance in the calcium metabolism probably has a contributory effect. Some investigators (BARSONY and KOPPENSTEIN, LYON and others) have observed cases with calcifications which presented certain signs which they regarded as being caused by the calcifications (»calcinosis intervertebralis» — »discite calcifiante»). SCHMORL, on the contrary, in common with HOLFELDER (cit. SCHMORL), considers the calcifications to have no clinical significance whatever.

The object of this treatise is to attempt to make a contribution to the etiology of these disc calcifications. It is based on observations both of calcifications elsewhere in the soft tissues of the body and of disc

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calcifications in three of the author's own cases. The treatise is also intended to show that these disc calcifications may be signs of a distinct disease entity. They may thus be of clinical significance from the differential diagnostic point of view.

A Survey of Roentgenologically Demonstrable Pathological Calcifications in the Soft Tissues of the Body

Calcifications in the soft tissues of the body are common discoveries during roentgen examinations: at the same time foci even though containing considerable quantities of calcium may not be demonstrated roentgenologically (Bloch). In lime deposits which are not too old, no ossifying cells may be seen microscopically within the calcified area, which is made up of more or less confluent clumps and masses of lime salts. The calcifications may thus present various appearances and sizes. From the differential diagnostic point of view it is important to remember that the roentgenogram obtained will not show any bony structure. In older lime deposits, single osteoblasts, arising secondarily, may sometimes be seen on microscopic examination. These latter may continue to grow, with gradual ossification of the entire lime deposit as a result. A roentgen examination of such ossified deposits may show a distinct bony structure by which they may be distinguished from ordinary calcifications.

With regard to the manner in which the lime deposits arise, investigators have for long drawn a distinction based upon a pathological differentiation between the following two forms:

1. *Metastatic Calcification.* — This is due to a super-saturation of the colloid solution which makes up the fluids. The histology of metastatic calcification is characterized by the normal appearance of the cells of the calcified areas in the early stages.

2. *Dystrophic Calcification* is obviously a purely local phenomenon. It seems to occur only in dead tissue, as a result of the physical and chemical alterations which take place therein.

It is not possible to determine from the roentgenogram whether the calcification is of a metastatic or dystrophic type. Certain calcifications, such as those occurring in a healed primary lesion in the lungs, pleura, or pericardium, as a result of an inflammatory process, or in the vessel walls in cases of arteriosclerosis, we know from experience to be for the most part dystrophic; we cannot however prove this roentgenologically. The method of determination in these cases consists of pathological examinations of the lime deposit and surrounding regions, although unfortunately it is only in exceptional instances that such examinations may be made by biopsy.

This classification into metastatic and dystrophic calcifications is based wholly on microscopic observations of the structure of the cells. This morphological examination gives no information as to their function, and thus does not show whether the latter is disturbed or not. In metastatic calcification there may be functional disturbances of a chemico-physical nature, occurring in cells apparently microscopically normal, which can give rise to lime deposits. It is probable that all calcifications — including the metastatic — are really a of »dystrophic» type, induced by a toxic agent.

The author feels, therefore, that the above classification into metastatic and dystrophic calcifications should no longer be used. He proposes another classification based on the *permanence* of the calcifications, and may be determined roentgenologically by control examinations. This basis of classification not only seems to be appropriate as regards appraisement of the individual case, but also forms a basis for more detailed study of the deep and essential difference between reversible and non-reversible calcification processes. The impermanent calcifications in particular present an extremely interesting field for research. It may be assumed that a more detailed and systematic study of conditions such as these will contribute to our understanding of the mechanism by which calcifications both develop and disappear, and thus clarify the etiology of these diseases.

Classification of the Pathologic Calcifications in the Soft Tissues of the Body, According to the Author

(Calcifications in the Secretory Glands and Tumours excluded.)

Group I. Permanent Calcifications.

1. *Calcifications in various Cartilaginous Tissues.*

a) In hyaline cartilage (FALCONER): in the costal cartilages, the sternal joints, and the xiphoid process; in the nose, larynx, trachea, bronchi; in the joints (joint cartilage) and the cartilaginous plates of the intervertebral discs (see below).

The author has made control examinations of 80 cases of costal calcifications observed in roentgen examinations of the lungs, these were carried out at intervals of from 2—20 years. In all these cases calcifications were permanent.

b) In fibrocartilage, found in the articular discs, menisci of the knee joint, and the peripheral part of the intervertebral discs (annulus fibrosus — see below).

c) Elastic cartilage in the ear, in the pharyngo-tympanic tube and in some of the cartilages of the larynx.

2. *Calcifications in the Vessels.*

a) In arteriosclerosis and syphilis;

b) In toxoplasmosis;

c) In thromboses of earlier date.

3. a) *Calcifications in the Lungs, Lymphatic Glands, and Serous Membranes in Tuberculosis.*

That these are of a permanent nature is clear from the following: The author has carried out follow-up examinations of 110 cases of calcifications in the lungs and pleura; these were examined roentgenologically twice, or often more, during an observation period of 10—20 years. In none of these cases was regression or disappearance of the calcifications observed. The same observations were made in cases with calcified lymph shadows in the abdomen. In 70 such cases the calcifications were unchanged in follow-up examination 5—18 years after the first examination.

b) *Calcifications in the Lungs and Hilar Glands* in Coccidiomycosis, in Histoplasmosis, and also in Blastomycosis, Aspergillosis, Moniliosis and possibly many other Mycoses. (DOUB.)

4. *Calcifications in the Connective Tissue.*

a) In Calcinosis Circumscripta.

b) In Calcinosis Universalis. This term has been reserved for cases which show more extensive calcification than in Calcinosis Circumscripta. Undoubtedly the cases of extensive calcification in elderly people, published by G. MOBERG, belong to this group. Both of these show a relationship to scleroderma and Raynaud's disease.

c) Lipocalcinogranulomatose (TEUTSCHLÄNDER, ANDREAS).

d) In hyperparathyroidism (in the eye, kidneys and elsewhere).

e) In certain rare diseases such as renal osteitis fibrosa cystica (ALBRIGHT and collaborators, LEVIN and GENOVESE), and in so-called pseudohypoparathyroidism (BROWNE).

f) Of hitherto unknown etiology in the dura mater of the brain.

5. *Calcifications in the Heart Muscle.* (GORE and ARONS.)

6. *Calcification in the Brain* in hypoparathyroidism (EATON and HAINES) and toxoplasmosis.

Group II. Impermanent Calcifications.

This group includes calcifications in the connective tissue resorbed spontaneously or as a result of therapeutic measures, such as roentgen treatment.

1. *Calcifications induced by toxic doses of Vitamin D.*

MULLIGAN and STRICKER have induced calcifications in different organs in dogs by the administration of large doses of Vitamin D. Even

in human beings extensive calcifications have been induced by such toxic doses. (BAUER and FREYBERG, HAM.) FREEMAN and associates have published roentgenograms of a man 49 years of age who had received large doses of Vitamin D. These show enormous calcifications in the vicinity of the shoulder and hip-joints which diminished or disappeared entirely when the large doses of Vitamin D were discontinued. Thus, to judge by this case, calcification processes of this type are reversible.

2. *Calcifications in Tendons and Muscles by Myotendinitis Calcarea.* Here we are dealing with a disease *sui generis*, with its own characteristic symptom picture previously described by the author. The pathological changes in this disease are found on the one hand, in the area where the tendons are attached to the bone and in the tendinous tissue in ligaments and articular capsules, and on the other, sometimes more pronounced, in the scanty connective tissue surrounding these tissues. Adjoining muscular tissue also shows similar changes. The histological features consist of alterative, proliferative and exudative changes, all the signs of inflammation. (SANDSTRÖM and WAHLGREN.) Since these calcifications were usually encountered outside the tendons in the peritendinous connective tissue, the author proposed that the disease be called *peritendinitis calcarea*. Subsequent pathological-anatomical investigations by the author have shown that in some cases the changes in the musculature in the vicinity of the tendon are strongly pronounced and more common than might be supposed on the basis of the location of the calcifications in the roentgenograms. Taking the localisation of the pathological changes into consideration the author considers the name *myotendinitis calcarea* more appropriate. In cases where the disease is localized exclusively in the tendinous tissue, such as the intervertebral discs, the name *tendinitis calcarea* may be used.

The disease is most common in the neighbourhood of the shoulder joints, particularly in the tendon of the supraspinatus and its adjoining muscular tissue, and in the hip-joints in the tendons of the greater and lesser trochanters. According to the author, other not uncommon locations are the vicinity of the elbow-joint, the knee-joint, the pelvic tendons, and the area surrounding the interphalangeal joints of fingers and toes. Another location (as this article is intended to show) is the central part of the intervertebral discs.

3. *Calcifications of unknown etiology in the intervertebral discs in childhood.*

Six such cases are published (WEENS a. o.), all characterized by an acute episode of pain, limitation of motion, spinal deformity and rapid change in the size of calcification. The etiology of the disease is still unknown. Myotendinitis calcarea may be a possible cause.

The above classification of calcifications into permanent and impermanent groups is not to be regarded as complete or definitive, but may undoubtedly be developed further. This applies to both the categories. Moreover, it is not improbable that one or more of the calcifications listed as permanent may, in the future, come under the heading of impermanent. A more thorough control might show that the calcifications in, for instance, parathyroidal disturbances, disappear following successful treatment of the basic disease. It may also be expected that by means of newer therapeutic measures in the future some of the permanent calcifications may be caused to resolve.

Calcifications in the Intervertebral Discs and their Relationship to the Anatomic Structure of the Discs

In the above classification, calcifications in the intervertebral discs have been listed as occurring in both the permanent and the impermanent groups. The explanation of this is that the intervertebral discs are formed by different tissues, and that the calcifications may occur in all of them.

According to FICK, SCHMORL, and others, they consist of three anatomically different parts:

1. A thin covering cartilaginous plate, demarcating above and below;
2. The peripheral, ring-shaped annulus fibrosus; and
3. The central nucleus pulposus.

The cartilaginous plates can become calcified like all other hyaline cartilaginous tissue in the body. These calcifications are of a permanent nature.

The annulus fibrosus is made up of fibrocartilage, which can also become calcified. According to RATHCKE's pathologico-anatomic investigations, calcifications of the discs are often found in this situation. He states that from one to several lime foci may be encountered, irregularly deposited over the annulus in a single disc. These calcifications are permanent in type, similar to fibrocartilaginous calcifications elsewhere in the body.

The more centrally located portion of the annulus, situated at the periphery of the nucleus pulposus shows, however, a structure which is microscopically identical to that found in ordinary tendinous tissue. (Fig. 1.) Any disease characterized by calcification, which can occur in fibrous connective tissue elsewhere, may be expected to occur in this part of the disc. The anatomy of the central part of the annulus permits the occurrence of both permanent and impermanent calcification. The

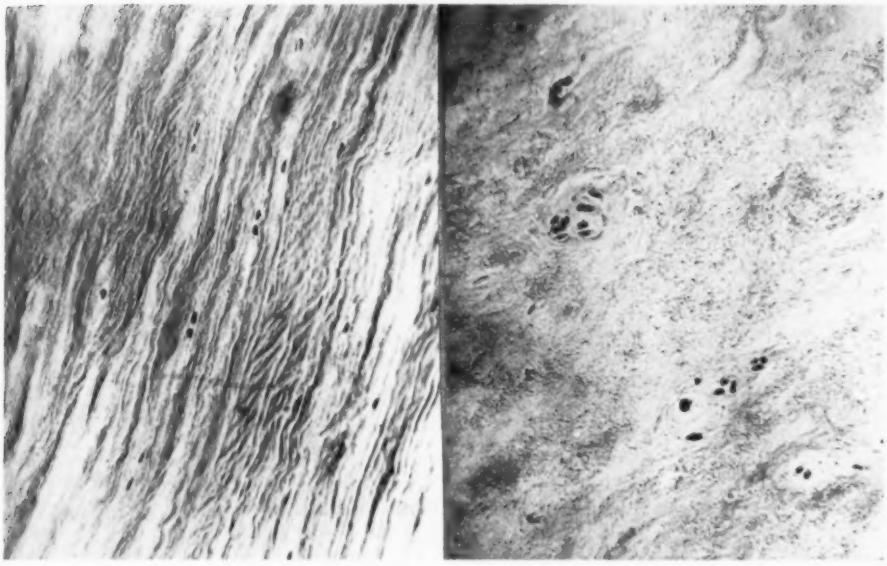

Fig. 1. Fig. 2.

Fig. 1. Central part of the annulus fibrosus showing arrangement of the collagenous fiber bundles similar to that in tendons. Magnification 190 ×.

Fig. 2. From the center of an intervertebral disc (nucleus pulposus). The collagenous tissue is sparse and irregular, corresponding to the arrangement in loose connective tissue. Magnification 190 ×. (From a 17-year-old boy.)

nucleus pulposus, which lies eccentrically in the annulus fibrosus and usually on the border between the medial and posterior third of the disc, forms a hollow space of irregular shape, often with a slight bulge forwards and backwards. Microscopically, the tissue here consists of thin sparse connective tissue. Fig. 2 shows that this connective tissue has the same morphologic appearance and structure as soft connective tissue elsewhere in the body, as for example in the peritendinous tissue.

Thus, conditions permitting the occurrence of both permanent and impermanent calcifications have been found in the nucleus pulposus. In consequence of the anatomical structure of the intervertebral discs the calcifications may occur as follows.

In the peripheral section of the discs (the cartilaginous plates, as in the annulus fibrosus) there may be permanent calcifications only and in the central part of the discs (nucleus pulposus and the adjoining

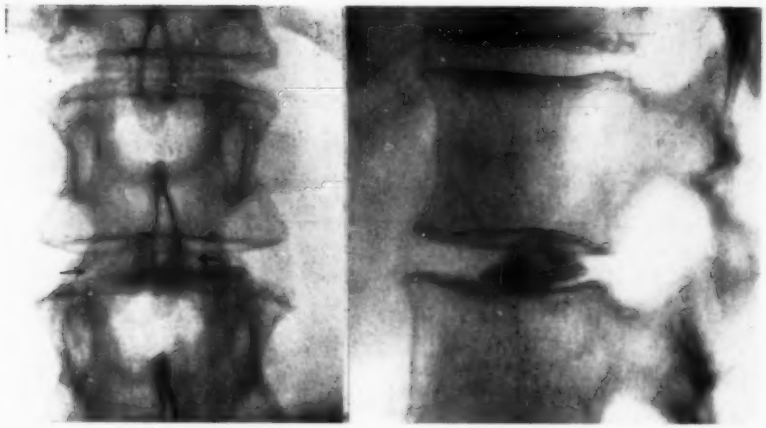

Fig. 3 a. Fig. 3 b.

Fig. 3 (a and b). Case I. 59-year-old woman with large calcifications corresponding to the central part of the first lumbar disc.

portion of the annulus fibrosus) both permanent and impermanent calcifications.

Concerning the clinical significance of these calcifications, the following should be observed:

The determination of impermanent calcifications in a disc, as elsewhere in the body, points to calcifications caused by toxic doses of Vitamin D or by *myotendinitis calcarea*. Hence such calcifications reveal a certain disease condition, and are of differential diagnostic significance.

As far as we know at present, permanent calcifications or at least those situated within the intervertebral plates, are without clinical significance.

Case Reports

Cases of Disc Calcifications of Impermanent Type

Case I. B. ♀, aged 59. About ten years previously a diseased condition resembling *myotendinitis calcarea* in the left shoulder; otherwise always healthy. No known trauma. A few weeks previously severe pains across the lumbar region, which subsequently increased. Pains intense; most pronounced upon getting up. The patient also had pain when lying down, particularly if she twisted her body. Temperature of up to 38.5 C during the two days previous to examination; had not received any extra Vitamin D.

Examination. Back very stiff; the patient could bend forward only slightly. Tenderness over the back musculature, which was severely strained. The patient noticed

Fig. 4 a. Fig. 4 b.

Fig. 4 (a and b). No calcifications in the disc.

direct and indirect sensitivity to pressure over the uppermost part of the lumbar spine. There was nothing noteworthy in the internal organs. S. R. (according to Westergren) 25 m/1 hr.

She was referred to the Roentgen Department with the diagnosis of acute lumbago and suspected acute spondylitis.

Roentgen examination. Two calcifications closely located and together the size of a coffee bean were seen in the disc between LI and LII in a position corresponding to that of the nucleus pulposus (Fig. 3 a). The lower and larger calcification was shaped like a bowl in which two smaller lime deposits seemed to be resting. Slight deforming alterations were observed in the adjoining vertebrae. On the wall of the abdominal aorta quite extensive calcification was noted (arteriosclerosis). On the anterior-posterior roentgenogram the calcification looked like a broad plate tapering off somewhat on the right side.

A centrally located pathological disc calcification, probably of impermanent nature and caused by (myo)tendinitis calcarea, was diagnosed.

Roentgen treatment was begun. The patient received 1×150 r every other day in five doses (size of field 20×20 cm, 180 kw, filter 1 aluminium $+ \frac{1}{2}$ mm Cu). She rapidly improved, and after only 20 days was free from discomfort.

Roentgen examination showed a slight decrease in calcification. The S. R. had now sunk to 10 mm/1 hr.

After 8 months a follow-up examination was made. The patient was subjectively and objectively free from signs. The roentgen examination showed that the calcifications previously observed in the disc between LI and LII had entirely disappeared. (Figs. 4 a and b.)

Case II. M. H. ♂, accountant, aged 50. For some time pains in the upper part of the back, whether lying down or erect. At times these pains radiated forwards around the chest walls. The patient had not taken extra Vitamin D.

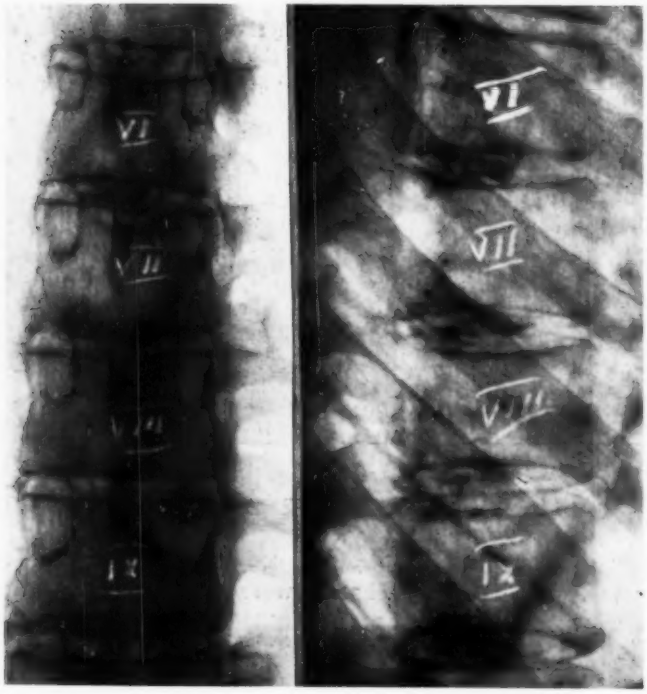

Fig. 5 a. Fig. 5 b.

Fig. 5 (a and b). Case II. 50-year-old man with calcifications in the central parts of the dorsal discs.

Upon examination a slight stiffness on movement in the upper spine was noted, and the patient complained of some pain. There was no direct or indirect sensitivity to pressure over the spine. S. R. 5 mm/1 hr.

Roentgen examination of the spine revealed a conglomeration of close-set, thin calcifications forming a plate centrally and somewhat posteriorly in the disc between the sixth and seventh dorsal vertebrae (Fig. 5 a). There was also a calcification in the disc between the seventh and eighth dorsal vertebrae which extended somewhat further forward in the disc than in the former case. Likewise, there was a smaller calcification in the disc between the eighth and ninth dorsal vertebrae. Fig. 5 b (A.-P.-view) shows the central position of the calcifications in the three intervertebral discs. The roentgen diagnosis was as follows: central calcifications, presumably impermanent and caused by (myo)tendinitis calcarea.

Roentgen treatment was begun. The patient received a dose of 150 r × 4 every other day (220 kw), size of field 10 × 20, $^1/_2$ mm copper and 1 mm aluminium, repeated after three weeks. After six weeks the patient was free from symptoms. A follow-up examination, which could no be made until six months later (Figs. 6 a and b) showed

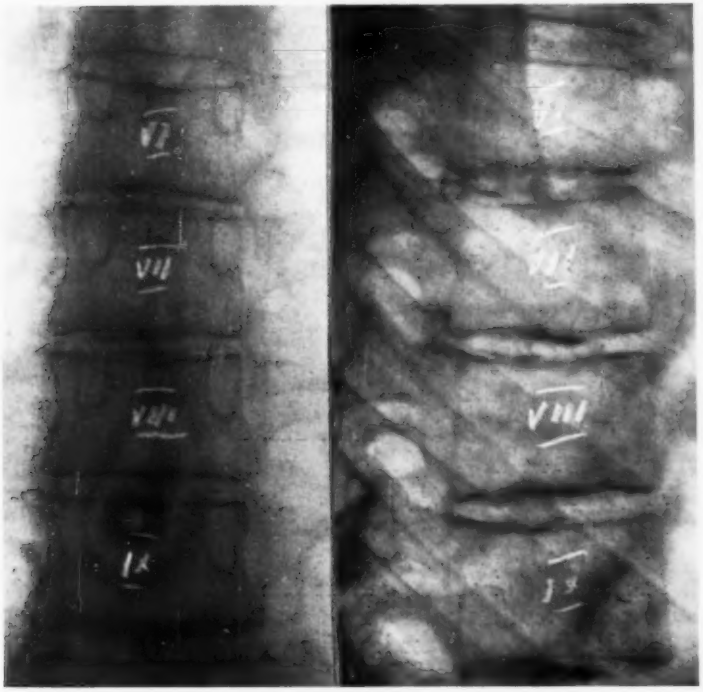

Fig. 6 a. Fig. 6 b.

Fig. 6 (a and b). Case II. Six months later. Distinct reduction or disappearance of the calcifications.

a considerable reduction in the calcifications in the three discs. Thus there were only a few small remnants in the disc between the sixth and seventh dorsal vertebrae. In the disc between the seventh and eighth dorsal vertebrae two wedge-shaped calcifications located nearly 1 mm apart, one above the other, the uppermost somewhat broader than the lower, persisted. (The calcifications may have been located within the peripheral part of the nucleus pulposus, where it adjoins the annulus fibrosus, or in the central part of the latter.) The calcification in the disc between the eighth and ninth dorsal vertebrae was completely resorbed.

Both these cases displayed central calcifications in the intervertebral discs. Some of them disappeared entirely, while others became smaller after roentgen treatment. Thus they may be classified as impermanent. There was no question of overdosage with Vitamin D. The only other cause of impermanent calcifications is the disease of the connective tissue and muscles which the author designates myotendinitis calcarea.

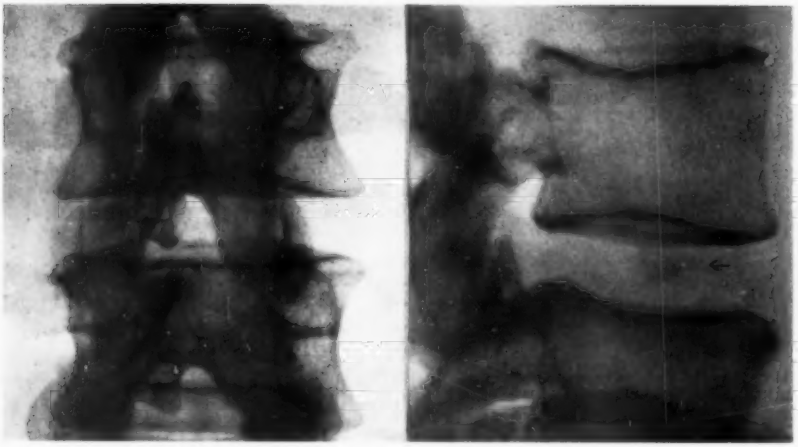

Fig. 7 a. Fig. 7 b.

Fig. 7 (a and b). Case III. A man, 54 years old, with calcifications of permanent type in the annulus fibrosus.

From the roentgenological, as well as the clinical point of view, the symptom picture and its course in these two cases correspond closely to those of this disease in other locations. Thus Case I is typical of the acute form of the disease, with acute initial pains increasing rapidly in intensity. Moreover, the patient displayed the general signs which are often encountered in acute cases, namely, a rise in temperature and an elevated sedimentation rate. By reason of the location of the changes deep within the body, however, the local signs which one often finds in, for instance, the shoulder and the hip-joints were less marked in this case. Thus there was no local swelling and distinct sensitivity to pressure. There was direct and indirect sensitivity to pressure with limitation of movement in the corresponding part of the spine.

Case II may be considered to typify the chronic form of the disease, with more prolonged and less subjective discomfort. Even the objective signs were not pronounced, except for the roentgenologically demonstrable calcifications which disappeared or were reduced in size.

Case of Disc Calcifications of Permanent Type

The following case from the author's material covering permanent calcifications has been included here in being typical and emphasizing the differential diagnostic questions which arise in connection with such cases.

Case III. C. E. R. E. ♂, aged 54. Pain in the back which started after a game of tennis in April, 1950, gradually becoming worse with occasional remissions.

The patient had had typical symptoms of lumbago and ischias with continual stiffness in the back and pain down the left sciatic nerve. Various forms of treatment, such as short wave, diathermy and immobilisation in plaster of Paris gave no relief.

Roentgen examination 26/6/50: Slight osteophytic outgrowths in the regions of anterior superior margins of L4—L5. Otherwise no alteration of the vertebrae. The discs are of normal thickness. In the disc between L3 and L4 a calcification slightly larger than a pea is seen. On account of acute and severe lumbago, the patient was admitted to hospital on 14/11/50.

Clinical examination of the patient suggested a disc hernia. There was also a markedly poor reflex of the left Achilles tendon.

Roentgen examination 18/11/50: The dense calcium deposit in the disc between L III and L IV demonstrated during the previous roentgen examination remains unchanged in size and density. Special films of the disc show that the calcification is situated in its right lateral part. This is clearly a case of calcification within the peripheral region of the disc (annulus fibrosus). The calcification is probably without clinical significance.

Myelography 22/11/50 showed the presence of a disc hernia between L IV and L V, which dislocated the root sheath in a curve. No alterations, however, could be noted in the neighbourhood of the disc (L III—L IV) where the calcification was present.

After operative removal of the disc hernia the patient became free from symptoms.

Roentgen-diagnostic Considerations Regarding Disc Calcifications

As the author has tried to show above, the occurrence of calcifications is possible in all parts of the intervertebral discs. In the peripheral parts, *i. e.* within the disc plates and the annulus fibrosus, the calcifications are of a permanent type. In the central parts of the disc, *i. e.* the nucleus pulposus and adjoining part of the annulus fibrosus, the calcifications may be of either permanent or impermanent type. It is, of course, impossible to determine with exactitude the part of the disc to which a calcification belongs; it must suffice if we can decide whether it is centrally or peripherally located, by trying to establish its position in relation to adjoining vertebrae. In order to do this, accurately centred, special roentgenograms of the calcified disc should be taken in two planes at right angles to one another, with the central ray directed over the disc in question. In determining whether or not the calcification is nuclear, one frequently finds confirmation in the fact that it has the form of a cast of the central hollow space and often a small protrusion forwards and backwards. If a calcification appears to be unusually large, and larger than the nucleus pulposus, it is probable that even the adjoining area of the annulus fibrosus forms part of the calcified area.

Peripheral calcifications of the disc located in the annulus fibrosus and the cartilaginous plates are, judging by the author's series of cases, somewhat rarer than the central. Differential diagnostic difficulties can here arise with regard to calcifications and ossifications occurring out-

side the disc in the surrounding ligament, as in the case of osteophytic outgrowths at the margins of the vertebrae, and so-called persistent epiphyses of the spine. In these last-named cases, however, a distinct bony structure is usually seen which makes a diagnosis possible. Nevertheless, for accurate diagnosis in these cases, roentgenograms should be taken in various planes and directions together with tangential films of the disc in question.

Calcifications in the cartilaginous disc plates have been included mainly on theoretical grounds. The plates are so thin that calcifications in them doubtless fully fail to appear on the roentgenogram owing to their close situation to the body of the vertebra.

All disc calcifications should be controlled by one or more examinations to decide whether they are of a permanent nature or not. This applies particularly to the central calcifications which can, as has been shown here, disappear. If, in a particular case, there is suspicion of the acute form of (myo)tendinitis calcarea, a follow-up examination should be made within a few weeks and repeated once or more often thereafter.

Clinical Considerations Regarding Disc Calcifications

Should a roentgen examination show the disc calcifications to be located within the *peripheral* part of the disc, they are probably, according to the above, of a permanent nature. These calcifications in the cartilaginous part of the disc undoubtedly occur in the same manner and for the same reasons as those which are seen in the cartilaginous tissue elsewhere in the body. As far as we know at present, these calcifications are of no clinical significance.

If, however, *centrally* situated disc calcifications are found, they may be either of permanent or impermanent type, and this cannot be determined by a single examination. In a case in which central calcifications have been found, a tentative diagnosis of myotendinitis calcarea is justifiable until diagnosis has been confirmed through roentgenological control examination. Other symptoms and signs of this disease may be present. The diagnosis of myotendinitis calcarea is the more probable if the patient has, or has had, signs of this disease elsewhere in the body earlier. In the author's long series of similar cases a large number of patients show signs of one or more relapses in various parts of the body. Hence in recording the patient's case history one should make particular enquiry concerning previous similar attacks.

If it can be shown that the patient has received large doses of Vitamin D, one would be equally justified in diagnosing an impermanent calcification. If, on the other hand, there are no reasons for assuming the existence of such a type of calcification, one must consider the

probability of a permanent calcification, and make a general examination of the patient with this in view. It must first be decided whether there be local or general calcinosis or a parathyreoidea disturbance. An investigation of the blood calcium should be carried out and a roentgenological examination made of other parts of the soft tissues of the body, in order to determine the presence of further lime deposits.

SUMMARY

The author proposes classifying pathological calcifications in the soft tissues of the body under two main headings, permanent and impermanent and gives examples of each. He shows that the anatomical conditions of the intervertebral discs permit the occurrence of both types of calcification: in the central parts, both permanent and impermanent, and in the peripheral parts, permanent types may exist. The author presents two cases of impermanent disc calcifications which were probably caused by the connective tissue and muscle disease which he calls myotendinitis calcarea, and one case with permanent calcifications without clinical significance. Hence these calcifications may be of clinical significance both from the differential diagnostic and the therapeutic point of view.

ZUSAMMENFASSUNG

Der Verfasser schlägt vor, die pathologischen Verkalkungen in den Weichteilen des Körpergewebes unter die 2 Hauptgruppen: — beständig und unbeständig — einzuordnen und bringt Beispiele dafür. Er zeigt, dass die anatomischen Verhältnisse der Intervertebralscheiben das Auftreten beider Verkalkungstypen zulassen: in den zentral gelegenen Abschnitten können beständige und unbeständige und in den peripher gelegenen Anteilen nur beständige Formen vorkommen. Der Verfasser stellt 2 Fälle mit unbeständigen Discusverkalkungen vor, die wahrscheinlich durch die von ihm »myotendinitis calcarea» benannte Bindegewebs- und Muskelerkrankung bedingt waren und einen Fall mit beständigen Verkalkungen ohne jedes klinische Krankheitszeichem von diesen. Somit können diese Verkalkungen im Hinblick auf die Differentialdiagnose und auf die Therapie klinisch bedeutungsvoll sein.

RÉSUMÉ

L'auteur propose de diviser les calcifications pathologiques de la chair en deux catégories principales: permanentes et non permanentes et donne des exemples. Il montre que les conditions anatomiques qui regnent dans les disques intervertébraux permettent l'apparition des deux types de calcifications: permanentes et non permanentes dans le centre, et, uniquement permanentes dans la périphérie. L'auteur présente deux cas de calcifications non permanentes au niveau des disques qui furent probablement suscitées par une affection du tissu connectif et des muscles qu'il nomme myotendinite calcaire. Il relate aussi un cas avec des calcifications permanentes sans symptôme clinique signifiant celles-là. Ces calcifications peuvent ainsi avoir de l'intérêt en clinique tant au point de vue diagnostic qu'au point de vue traitement.

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FROM THE ROENTGEN DEPARTMENT (DIRECTOR: PROFESSOR ETTORE DE BERNARDI)
OF THE ST. FRANCESCO HOSPITAL, NUORO, ITALY

PULMONARY HYDATID DISEASE IN MAN[1]

by

Ettore de Bernardi

The following short description of certain changes occurring in the lungs in human echinococcal infection may be of value to those radiologists unfamiliar with the disease. Hydatid disease is quite unknown in many countries but in certain others *e. g.* Iceland, Australia, and Italy, its frequency is such that the disease is considered endemic. In Italy it often occurs in the area around Rome, in the Abruzzi, and even more often in Sicily, Sardinia and the province of Grosseto; in other parts of the country the disease is almost unknown.

I shall not discuss the pathogenic theories nor the frequency of the situations of the disease (in the liver, lungs, bones &c.) and the way in which they vary in different countries, but shall confine myself to the radiological pulmonary changes only. A radiological diagnosis is undoubtedly possible even in the early stages of the disease. This is especially true in the pulmonary region where the cyst may be detected in the initial stages when a clinical diagnosis is impossible.

The case shown in fig. 1 is that of a man aged 64 who was examined for a suspected rib fracture. The roentgen examination revealed a typical case of hydatid disease of the lungs. Two cysts of almost equal size can be seen near the lower part of the left hilum: they are circular in shape with the outlines sharply defined. There was good movement of the diaphragm. The discovery was somewhat surprising as the patient had no previous pulmonary history. Another example is shown in fig. 2 of a man aged 34 with an hydatid cyst of the right lung. The roentgen film shows a typical sharply circumscribed round shadow of moderate density and without peripheral reaction lying in the lateral region of the right lung. A small less opaque but clearly defined shadow is seen on the left side in the inferior part of the hilum: it contains a three-lobed nucleus indicating calcareous degeneration of the contents of the cyst.

The shape of pulmonary hydatid cysts depends upon their manner of growth and tends to be spherical. According to many writers, the characteristic shape of the cysts is rounded, whilst others maintain that

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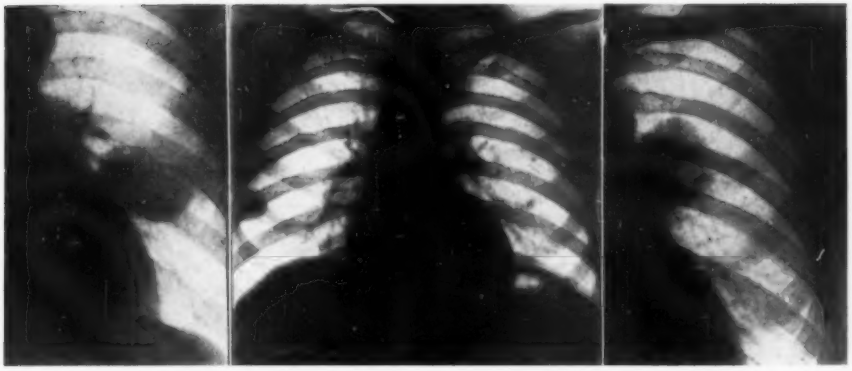

Fig. 1. Fig. 2. Fig. 3.

Fig. 1. Two circular shadows with well-defined margins are present in the middle zone of the left lung

Fig. 2. A small round cyst lies in the lateral region of the right lower zone. A smaller less opaque cyst with a calcified nucleus lies in the region of the left hilum.

Fig. 3. A single well-defined cyst protruding from the upper part of the left hilum.

it is oval. The direction of development of the cyst and its final form depends upon its situation and the contiguous anatomical structures. Thus it is only a cyst centrally situated which will in all probability remain round (fig. 3). When there are multiple cyst formations, the normal rounded shape may be changed by pressure from neighbouring cysts. In such cases, fusion is prevented by the strength and resistance offered by the external laminated cuticle and to a lesser degree by the enveloping membrane. When there is no resistance, however, the spherical form is maintained. In fig. 4, we have an example of these multiple cysts. A very large cyst, certainly the oldest, lies close to the heart on the left side, and just above it a smaller and well-defined cyst is seen. These cysts are causing pressure upon one another and are consequently becoming deformed. An oval cyst lies close to the vertebral column. The upper cyst on the left side is almost empty and a little residue only remains in its lower part. This cyst which is lying free has a typical rounded appearance similar to the cyst visible at the right base. (At the right base there are two cysts, but only one of them is clearly visible in fig. 4.) In a further examination carried out sixteen months later (fig. 5), we see that the cysts have become enlarged to a considerable and uniform degree. The paravertebral cyst has become smaller and has now no characteristic appearance. The upper cyst which was seen to be almost empty at the previous examination, is now also much reduced in size

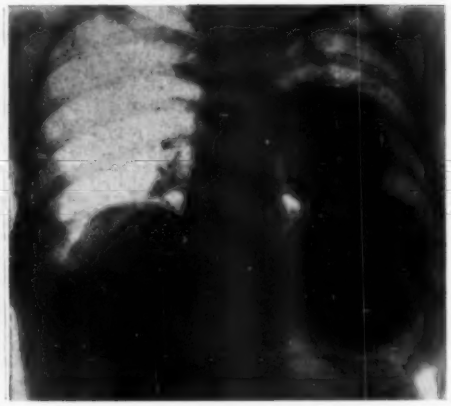

Fig. 4. Fig. 5.

Fig. 4. Three deformed cysts on the left side and one (the uppermost) spherical. The latter is almost empty. In the lower part a disjoined, collapsed membrane is seen. At the right base there are two cysts, but only one of them is visible in this figure.

Fig. 5. Same case, sixteen months later. The cysts on the left side have grown and are irregular in shape. The cyst in the left apical region is seen to be collapsed.

and appears as a network of slender lines directed upwards and outwards. An area of probable atelectasis has developed at the left apex. The cyst at the right base, having room for expansion, has grown more regularly and has maintained the characteristic appearance.

Not all authors agree on the characteristics which distinguish hydatid cysts. Opinions have slowly changed and the majority do not agree today with BECLÈRE's description of the pulmonary cyst as being round but consider it better described as oval. Originally the cyst is round but it will grow as its environment permits and its shape will thus be determined. Opinions also differ as to whether a cyst may be described as clearly defined or not. In my opinion the radiological appearances may vary in different cases due to the condition of the fibrous pericystic membrane which is normally very thin and only slowly grows thicker and less regular. The outline thus depends largely upon the age and size of the cyst and may be masked by atelectasis or reactive changes in the surrounding structures. The development of an echinococcal cyst is normally very slow and it may take years for it to reach the size of, say, an orange. In exceptional cases, it may become very large.

The case shown in fig. 6 is of a boy aged 15. The cyst which occupies almost the whole of the left lung presents an oval form owing to pressure, but the lower part is

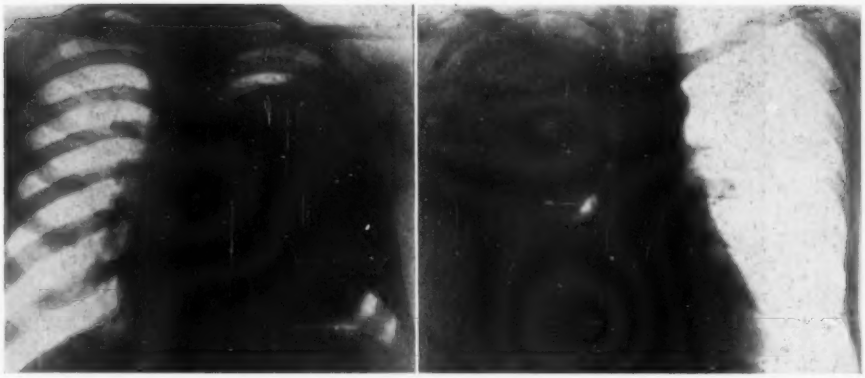

Fig. 6. Fig. 7.

Fig. 6. A large oval cyst occupies almost the whole of the left lung field.

Fig. 7. Large cyst in the right upper and middle zones. Below it a further cyst masks the diaphragm.

rounded and is clearly distinguished from the dome of the diaphragm. A differential diagnosis in cases such as these may be difficult particularly in eliminating cysts of another nature. In the case shown in fig. 7, one cyst occupies almost the entire right lung and below it a further cyst is present just above the diaphragm. The outer margin of the upper cyst is in intimate contact with the thoracic wall whilst the inner margin is clearly defined and separated from the mediastinum by a small border of lung tissue.

Difficulties in diagnosis may arise, not only by the size of the cyst but also by the mediastinum, the diaphragm or the chest wall masking its outline. We have an example of this in the case shown in fig. 8. The cyst lies at the base of the left lung and is masked by the left dome of the diaphragm. A gas bubble if present in the stomach may be of assistance in defining the left dome. A differential diagnosis from basal pleurisy may be made by noting that the upper margin of the opacity is convex, and that there is no evidence of marginal pleural thickening. The upper border, particularly in the central part, of the shadow presents a double outline. This signifies disjunction of the membrane, a pathognomonic sign of an echinococcal cyst, according to all authors. It may be referred to as the »disjoined membrane». A similar sign of equal importance, the sign of the »floating membrane» may also be said to be pathognomonic. In the case shown in figs. 4 and 5, an examination carried out two months after the patient had had a »vomica» showed that the uppermost cyst in the left lung was deformed and half filled with fluid, the upper part containing air. The irregular fluid level, the sign of the »floating membrane» is present (fig. 4). The round opaque mass

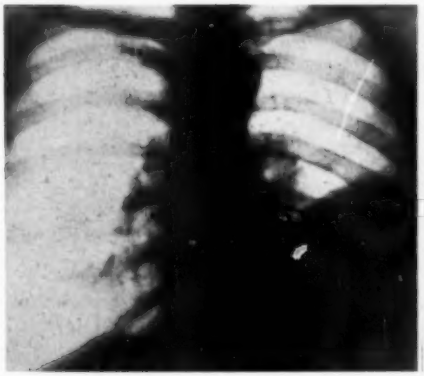

Fig. 8.

Fig. 9.

Fig. 8. Cyst lying at the base of the left lung masks the dome of the diaphragm. Note the convex upper margin.

Fig. 9. A large basal cyst partially air-filled with an irregular fluid level owing to the presence of a »floating» membrane. Above it, some floating cystic offshoots.

lying in the right lung possesses an irregular double upper margin and in the middle of this a fine translucent band may be detected. This is the sign of the »disjoined membrane» (fig. 5).

The next case (fig. 9) is that of a girl of twenty. The cyst was at first atypical and the diagnosis doubtful. The second roentgen examination carried out after a »vomica» provided a completely different picture, however, and showed a large cavity, the basal portion of which was opaque and the upper filled with air. The middle third of the fluid surface is seen to be irregular owing to the presence of the »floating membrane» and round, translucent cystic off-shoots. It is thus the growth and the changes in the cyst which sooner or later produce signs which may be said to be characteristic of hydatids. It may be here observed that the radiological diagnosis is not always confirmed by the Casoni or the Ghedini Weimberg serum reactions.

I have encountered cases with positive serum reactions and in which a roentgen examination indicated the presence of a tumour but operation revealed that the mass was not an echinococcal cyst. On the other hand a negative serum result has sometimes been obtained in the presence of typical roentgen changes. Such discrepancies however but seldom occur.

Conclusions

From this brief and summary presentation of the radiological picture of pulmonary hydatid disease, it may be gathered that the diagnosis is usually possible from the characteristic shape and outline of the cyst, always remembering that these depend upon its age and ability to spread. The situation of other growing cysts will of course determine the latter but expanding unhindered we believe that a cyst tends to be rounded rather than oval. It may be subsequently modified by pressure from other cysts or by surrounding structures. A hydatid cyst may present no symptoms in the very early stages so that its discovery by roentgenography may come as a surprise. If there be no restrictions or obstacles to its expansion, a cyst may not grow so slowly, however, and growth may take place at the expense of surrounding cysts which are diminishing in size or withering.

The ageing of a cyst is evidenced by the breaking down of the membrane which allows the contents to empty into the pleural cavity or bronchus. The cyst then takes the form of a cavity containing air and the pathognomonic signs of »disjoined membrane», »floating membrane» or cystic off-shoots above the surface of the cystic fluid, may appear. The death of a cyst is indicated by the appearance of a chalky membrane, a sign but seldom encountered. It is at the early stages that there may be difficulties in the differential diagnosis. Cysts of another etiology, lung abscess, benign and malignant tumours will all be considered. Conditions such as encysted pleurisy rarely present difficulties. It is as the cyst breaks down that the pathognomonic signs become apparent.

SUMMARY

The author discusses the roentgenological signs of pulmonary hydatid disease, which are important in making a differential diagnosis, and draws attention to the growth and modification in the appearances of the cysts. Some illustrative cases are presented.

ZUSAMMENFASSUNG

Verfasser erörtert radiologische Einzelheiten über Lungenechinococcus, die für die Differentialdiagnose besonders wichtig sind: pathognomonische Zeichen, Form und Evolution. Einige charakteristische Fälle werden gezeigt.

RÉSUMÉ

Dans les cas de kyste hydatique l'exploration radiologique donne des renseignements d'une importance capitale pour le diagnostic différentiel, avec des syndromes radiologiques charactéristiques. L'auteur présente quelques cas typiques.

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DE LA CLINIQUE RADIOTHÉRAPEUTIQUE (CHEF: PROF. DR. D. J. STEENHUIS)
DE L'HÔPITAL UNIVERSITAIRE DE LEYDE, HOLLANDE

APERÇU DU TRAITEMENT DES ANGIOMES DANS LA CLINIQUE RADIOTHÉRAPEUTIQUE DE L'HÔPITAL UNIVERSITAIRE DE LEYDE DE JANVIER 1946 JUSQU'EN MAI 1949 (308 CAS)[1]

par

P. J. L. Scholte

Les angiomes font partie du groupe des hématomes (DEELMAN). Certains d'entre eux doivent être traités pour des motifs esthétiques et d'autres parce qu'ils peuvent devenir gênants et menacer le bon fonctionnement de certains organes par leur croissance illimitée.

Au cours des 50 dernières années plusieures études ont été publiées se rapportant au traitement des hémangiomes au moyen des rayons de roentgen et du radium. Les résultats publiés sont favorables, surtout ceux obtenus avec le type caverneux et le type tubéreux.

La dose nécessaire pour obtenir un effet thérapeutique ne provoque pas de nécrose. Dans bien des cas cette dose est tellement petite qu'elle ne provoque de changements fibreux que dans les petits vaisseaux: ceux-ci s'oblitèrent alors de sorte qu'il ne subsiste même pas de cicatrice.

C'est donc une manière de guérir qui ressemble de très près à la guérison spontanée que l'on constate parfois. Une seule publication, celle de LITER (1936) rapporte les chances de guérison spontanée du type caverneux chez 77 malades: 50 % de ces tumeurs cessèrent de croître après un certain temps tandis que le reste disparût spontanément. D'autres (NANCEL, DENARDET et WINER) mentionnent des résultats analogues.

Notre propre expérience est moins heureuse, vu que dans les cas de traitements différés ou interrompus pour cause de maladie, nous dûmes constater une croissance nette des angiomes.

Le petit tableau suivant rapportant l'âge moyen des petits malades traités par nous est très démonstratif.

[1] Soumis à la Réd. le 10 mars 1951.

Tableau I

Formes superficielles	âge moyen	4.8 mois
Formes souscutanées	âge moyen	7.7 mois
Formes très étendues	âge moyen	16 mois
Formes multiples	âge moyen	3.7 mois

(Dans ces chiffres ne sont pas considérés les quelques cas adultes.)

Nous pensons donc devoir insister sur un traitement commençant aussi tôt que possible, éventuellement au cours du premier mois. Les chances de guérison sans cicatrice ni d'autres conséquences sont d'ailleurs d'autant plus grandes que l'enfant est plus petit, parce qu'à ce moment l'angiome est le plus radiosensible. Comme il n'est pas possible de prédire quels angiomes vont disparaître spontanément ou cesser de croître, il ne nous semble pas bon d'attendre; un traitement correct par contre n'est jamais défavorable et exclut tout risque ultérieur.

Groupes cliniques

Il est important de distinguer les types tubéreux et caverneux des autres formes cliniques insensibles au rayons.

Formes non sensibles

Parmi les formes non radiosensibles nous mentionnons le naevus flammeus ou tache de vin et la télangiectasie (spider naevus ou naevus araneus). Tant que ces formes sont jeunes et rouge clair il est possible d'obtenir des résultats avec une irradiation superficielle (contact, radium et rayons Bucky). La couche endothéliale des vaisseaux par contre n'est pas radiosensible; PENDERGRASS attribue cela au fait que l'endothélium est composé de cellules du type adulte. De plus les lymphangiomes et les formes mixtes, les angio-lymphangiomes exigent des doses très élevées.

Formes sensibles

Ce sont les naevi vasculaires hypertrophiques surélevés, nodulaires, parfois lobulés et mous (formes tubéreuses, MAC KEE parle de strawberry-marks). Ces formes peuvent s'étendre sous la peau et occupent un volume énorme (formes caverneuses).

Nous voyons souvent des taches rouge vif entourées d'une zone bleuâtre. Dans ces cas il s'agit plutôt de capillaires d'origine veineuse. Lorsque l'angiome est plus grand, étendu dans la profondeur et qu'il présente des pulsations, on parle d'angiome racémeux.

Matériel clinique

De janvier 1946 à juin 1949 nous avons traités 308 malades porteurs d'angiomes radiosensibles. On peut les subdiviser d'après le type de la tumeur:

Groupe I 50 %. 153 malades avaient une tumeur superficielle ne dépassant pas un diamètre de 2 cm et une épaisseur de 3 mm (forme tubéreuse).

Groupe II 20 %. 59 malades avaient une tumeur ne mesurant pas plus que 2 cm de diamètre et 3—5 mm d'épaisseur et entourée d'une zone bleuâtre (forme caverneuse).

Groupe III 14 %. 44 malades présentaient une tumeur dont le diamètre dépassait 2 cm et l'épaisseur plus de 5 mm, entourée d'une zone bleuâtre et souvent composée de bosselures confluentes (grosses formes caverneuses souscutanées).

Groupe IV 1 %. 6 malades présentaient une tumeur étendue menaçant la fonction de certains organes et dont une fut même incompatible avec la survie.

Groupe V 15 %. 46 malades étaient porteurs de tumeurs multiples. En général la multiplicité se voit dans les formes superficielles et petites, parfois combinées avec une ou plusieurs plus grandes, mais jamais avec des tumeurs du groupe III.

L'âge moyen des petits malades traités par nous fut de 3.7 mois, donc appartenant au groupe le plus jeune (tableau I). (Il est possible qu'un traitement précoce prévienne une croissance plus étendue.) La répartition sur le corps ne différa pas des tumeurs solitaires tandis que le nombre par malade fût très variable. Voici la répartition des cas selon la localisation de la tumeur:

Sur la tête ..... 38 %
(La face 28 % — front 9 %, autour des yeux 6 %, nez 3 %, joues 4 %, lèvres 3 %, muqueuse de la bouche 1 %, menton 2 %; cuir chevelu 8 %, oreilles 2 %)
Le cou et la nuque ..... 3 %
(le cou 2 %, la nuque 1 %)
Les extrémités superieures ..... 9 %
(bras 3 %, avant-bras 2 %, mains 4 %)
Tronc ..... 39 %
(poitrine 6 %, dos (partie sup.) 16 %, épaules 3 %, abdomen 5 %, dos (partie inf.) 2 %, hanches 7 %)
Organes génitaux ..... 3 %
Les extrémités inférieures ..... 8 %
(jambes 5 %, pieds 3 %)

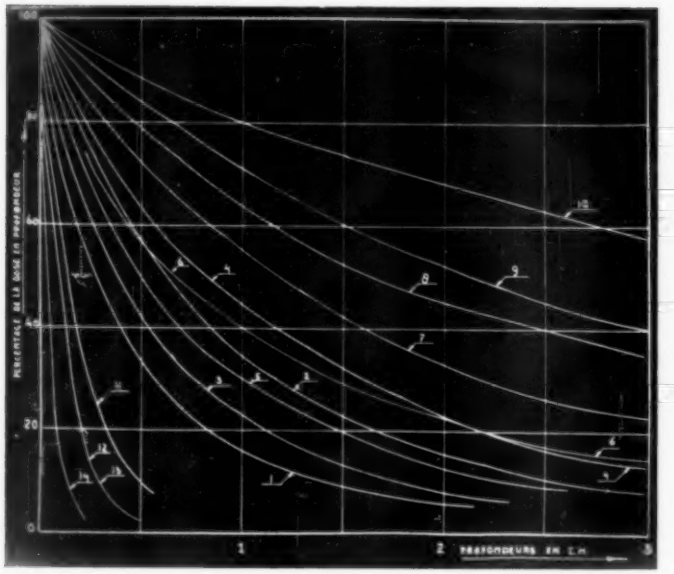

Fig. 1. Explication du graphique (composé par Dr. J. H. MELLINK).

1. Appareil à contact PHILIPS Hôpital académique de Leyde: 50 kV distance foyer —peau 20 mm. Filtre inhérent et philite.
2. Appareil à contact PHILIPS Hôpital académique de Leyde: 50 kV d. f. p. 20 mm. Filtre inhérent + 2.5 mm Al.
3. Appareil à contact PHILIPS Hôpital académique de Leyde: 50 kV d. f. p. 40 mm Filtre inhérent.
4. Appareil à contact PHILIPS Hôpital académique de Leyde: 50 kV d. f. p. 40 mm. Filtre inhérent + 2.5 mm Al.
5. Appareil à contact PHILIPS Hôpital académique de Leyde: 50 kV d. f. p. 110 mm. Filtre inhérent.
6. Rayons γ du Ra, filtrés par 0.5 mm Pt. Distance moulage—peau 1 cm.
7. Rayons γ du Ra, filtrés par 0.5 mm Pt. Distance moulage—peau 2 cm.
8. Appareil pour thérapie profonde Hôpital académique de Leyde 100 kV d. f. p. 30 cm. Filtre inhérent de 3 mm Al.
9. Appareil pour thérapie profonde Hôpital académique de Leyde 140 kV d. f. p. 30 cm. Filtre inhérent de 3 mm Al.
10. Appareil pour thérapie profonde Hôpital académique de Leyde 180 kV d. f. p. 30 cm. Filtre total de 0.5 mm Cu.
11. Contact de rayons β du Ra filtrés par 0.1 mm de monel (d'après BLOMFIELD & SPIERS).
12. Contact de rayons β du P_{32} (d'après LOW-BEER).
13. Rayons X de 25 kV d. f. p. 6 cm sans filtre (d'après EBBEHOJ).
14. Rayons X de 12 kV d. f. p. 9 cm sans filtre (d'après EBBEHOJ).

STRANDQUIST avait trouvé 10 % d'angiomes de plus sur la tête. Pour le reste, les chiffres suédois et américains ne diffèrent pas des nôtres (PENDERGRASS).

Certaines localisations exigent une méthode spécialement prudente d'irradiation:

le pourtour des yeux	3.5 % des cas
le cou	2 % des cas
l'abdomen et la partie inf. du dos	1.5 % des cas
la région mammaire	0.5 % des cas
les organes génitaux	2.8 % des cas
les pieds et les mains	5.8 % des cas

donc en tout 22.6 % des cas. Les difficultés qui se présentèrent dans ces cas seront décrites plus loin.

Nous n'avons pas vu de rapport entre la localisation d'une tumeur et son étendue. 7 % des angiomes présentaient d'emblée des ulcérations avant tout traitement. En général c'étaient des angiomes des plis de flexion (pli inguinal, cou, fesse). Quoique le traitement de ces cas ne soit pas contreindiqué, une guérison sans cicatrice est impossible.

Répartition selon le sexe et l'âge

36 % des cas étaient des garçons, 64 % des filles. STRANDQUIST avait trouvé chez ses 300 malades du Radium-Hemmet 27 % et 73 % respectivement. Il ne s'expliquait pas cette différence que l'on retrouve généralement dans la littérature. En Suède le nombre de naissance chez les garçons ne diffère pas de celui des filles, en Hollande il y a un peu plus de naissances mâles. La différence entre le nombre de cas chez les garçons et celui chez les filles doit être attribuée à la sollicitude plus grande des parents pour l'aspect de leurs filles. Cela semble d'écouler du tableau suivant:

Tableau II

Age lors du premier examen:

1— 3 mois	30 garçons et 67 filles.
3— 6 mois	31 garçons et 65 filles.
6—12 mois	19 garçons et 36 filles.
plus d'un an	21 garçons et 18 filles.

Les formes plus sérieuses qui sont la règle chez les groupes les plus âgés sont en effet aussi fréquentes chez les garçons que chez les filles, tandis que dans les groupes plus jeunes les parents semblent montrer une tendance à attendre une guérison spontanée, quand il s'agit de garçons.

Traitement

Nous ne nous bornons généralement pas à réduire ou inactiver l'angiome mais nous tâchons d'obtenir le résultat esthétique le meilleur possible. Notre ligne de conduite consistait en un choix exact de la qualité des rayons et un dosage aussi petit que possible. Dans le choix de la dureté des rayons nous tenons compte de l'étendue et de la profondeur de la tumeur.

Nous disposons à l'hôpital Académique de Leyde des modalités de rayonnement suivantes dont le graphique de la fig. 1 donne les courbes d'absorption et une explication.

Les rayons les plus durs (kilovoltage élevé) sont nécessaires lorsque la tumeur s'étend plus dans la profondeur, parce qu'ainsi l'absorption dans la profondeur est relativement plus élevée.

L'emploi du radium semble paradoxal. Ce dernier filtré au moyen de 0.5 mm Pt ne donne pratiquement que des rayons gamma qui correspondent au rayonnement que donnerait une source de rayons roentgen de 1.2 millions de volts. Le graphique (courbes 6 et 7) nous montre cependant que, différant en cela de la courbe 10 de l'absorption de rayons d'un appareil pour radiothérapie profonde de 180,000 volts, la courbe d'absorption du radium présente une pente raide. Celle-ci est causée par la petitesse de la distance entre le radium et l'objet irradié qui ne dépasse pas 2 cm. Il en résulte que les courbes 6 et 7 ressemblent à celles de l'appareil à contact de 50,000 volts à une distance de 4 cm et filtré par 2.5 mm Al (courbe 4). Les quantités de radium dont on dispose généralement ne permettent pas une téléradium-thérapie (télécuriethérapie).

Notre expérience personnelle et celle d'autres centres (4, 6, 7, 8) nous ont appris que l'irradiation au moyen de radium donne des résultats plus satisfaisants du point de vue esthétique. L'épiderme est moins lésé et les vilaines taches pigmentées que l'on voit souvent paraître après de fortes doses de rayons mous ne se voient pas lorsqu'on emploie des doses même élevées de radium. Mayneord et Edith Paterson citent à ce point de vue la radiation caractéristique moins intense de l'atome de soufre tandis que Gray mentionne une autre division en ions: cela conduirait trop loin d'en parler en détail ici. Nous préférons donc l'emploi de radium quand il s'agit d'angiomes de la face.

L'application des moulages de radium exige cependant une technique qui surtout chez les enfants très mobiles, peut présenter des difficultés.

Dernièrement, nous avons remplacé les moulages en cire par des moulages en feutre. (Fig. 2.) Ces derniers sont plus faciles à appliquer, la sparadrap ne se déplace pas de sorte que le système de dosage selon la méthode de Manchester devient plus facile à appliquer et plus correct.

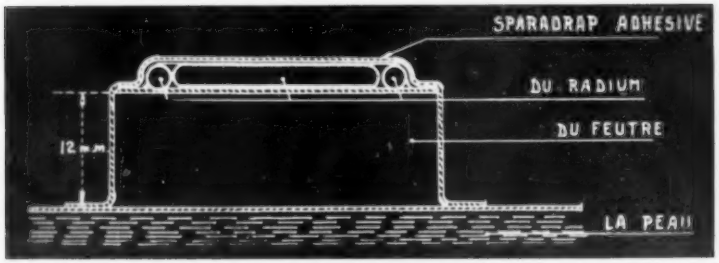

Fig. 2.

Cependant le feutre est légèrement compressible ce qui est un inconvénient; des expériences nous ont cependant montré que cette erreur est négligeable dans les dosages modérés. Lorsque nous employons du feutre de 1.2 cm d'épaisseur en tenant compte de l'épaisseur du sparadrap et de la demie-épaisseur du tube de radium nous pouvons utiliser sans grand inconvénient les dosages calculés pour une distance de 1 cm.

Pour un tel traitement, l'enfant doit être admis à l'hôpital, ce qui présente parfois des difficultés. Une autre manière d'utiliser le radium que nous employons souvent consiste en une implantation: des aiguilles contenant du radium sont implantées sous la peau dans la tumeur. La répartition est géométrique et tient compte des règles de répartition des rayons. De cette façon on atteint une dose suffisante dans la profondeur sans que le dosage à la surface soit excessif. Cette méthode peut présenter un intérêt tout particulier quand il s'agit de traiter des hémangiomes souscutanés profonds. Elle présente cependant un inconvénient: il est techniquement plus difficile d'obtenir ainsi une répartition exacte du radium que lorsqu'on emploie des moulages. Cependant, comme il s'agit ici de doses modérées, une technique même mauvaise ne doit pas provoquer de nécrose. Un procédé plus souple et plus exact consiste en l'emploi de »granules» de radon au lieu d'aiguilles de radium.

Tumeurs difficiles à irradier à cause de leur localisation

Lors de l'irradiation de tumeurs avoisinant l'orbite il faut absolument épargner le cristallin. Nous ne pouvons protéger les yeux au moyen d'œillères en plomb de 1 mm d'épaisseur que si nous utilisons des rayons mous. L'emploi de radium sous forme de moulage n'est donc pas à conseiller ici et l'emploi d'aiguilles interstitielles n'est permis que lorsqu'il s'avère impossible d'obtenir sans lésions cutanées un dosage suffisant dans la profondeur à l'aide de l'appareil à contact.

L'irradiation d'angiomes de la paume de la main et de la plante des pieds et, chez les filles, d'angiomes de l'abdomen et du cou doit littéralement rester limitée à la superficie, car là aussi le dosage dans la profondeur pourrait devenir nuisible. Il est évident que ceci peut parfois présenter des difficultés. Dans la plupart des cas il est cependant possible d'irradier une lésion située sur le scrotum (cas heureusement rare) en recouvrant les testicules d'une couche de plomb et en utilisant des rayons peu pénétrants.

Dans les cas où un traitement par les rayons présenteraient trop d'inconvénients il faudra choisir une autre méthode, mais ce choix ne sera pas facile: l'ablation chirurgicale peut constituer une opération d'envergure, l'application de neige carbonique ne fait disparaître que les macules les plus superficielles. Nous n'avons aucune expérience personnelle de l'emploi de liquides sclérosants mais on nous a envoyé maintes fois des malades chez qui un traitement de ce genre n'avait pas conduit au résultat désiré. L'application ultérieure de rayons chez de pareils malades aboutit généralement à une vilaine cicatrice, la peau ayant déjà été lésée.

Les soi-disants rayons Bucky (rayons roentgen de 10 kV) restent très superficiels (voir le graphique). A 1 mm sous la surface de la peau la dose tombe à un dixième de la dose à la superficie. Il est donc possible de provoquer une nécrose de la peau tandis que l'angiome pratiquement indemne continue à s'étendre dans la profondeur. Les parties rouges de l'angiome qui étaient superficielles et donnaient une impression d'activité sont détruites mais plus bas rien n'est changé. Les succès obtenus ainsi dans les traitements d'hémangiomes souscutanés sont dus je pense, à une tendance à la guérison spontanée. Quand celle-ci ne se produit pas, l'hémangiome qui d'abord aurait pu être traité avec facilité devient un problème radiothérapeutique. Une implantation de radium, une irradiation à kilovoltage élevé ou un moulage prolongé à 2 cm de distance restent seuls possibles si du moins la localisation le permet (p. ex. pas au poignet). Ces méthodes ont pour but d'obtenir un dosage aussi élevé que possible dans la profondeur en épargnant dans la mesure du possible, la peau déjà fortement lésée. Le résultat en est généralement une cicatrice plus ou moins laide. L'expérience nous a appris le fait remarquable que les angiomes cutanés semblent croître et s'étendre dans la profondeur lorsqu'ils sont exposés aux rayons Bucky mais nulle part dans la littérature nous avons trouvé une confirmation de ce phénomène. On a cependant décrit un œdème qui pourrait apparaître brusquement à la suite d'un dosage élevé de rayons Bucky (MACKEE), ce qui pourrait peut-être expliquer nos observations.

Dosages

Matériel clinique du groupe I

Ces angiomes superficiels guérissent bien avec le Philips-contact à 20 mm. On donne 1 à 3 doses de 620 r à 765 r espacées de $1^1/_2$ mois à 2 mois en utilisant des champs aussi petits que possible. Plus on a de patience plus le résultat est beau. Quand l'angiome ne croît plus et devient plus pâle il vaut mieux omettre une séance.

Quand l'angiome est situé à la face il est bon de donner une dose plus modérée par séance et de faire revenir l'enfant plus souvent. Chez presque tous les malades traités de la sorte la cicatrice ne présente pas ou presque pas de pigmentation et est pratiquement invisible. S'il existe déjà de la nécrose ou si celle-ci apparaît au cours du traitement à la suite d'un traumatisme ou pour une autre raison, une cicatrice plus visible est inévitable. Celle-ci ressemble alors à une cicatrice postvaccinale. Il ne semble pas nécessaire de traiter ce groupe au moyen de radium.

Les taches situées sur les lèvres et la muqueuse buccale sont moins radiosensibles. On utilise donc pour ces dernières des doses de 765 r par séance.

Les dosages décrits plus haut peuvent au niveau du cuir chevelu, provoquer une épilation temporaire.

Dans les formes très superficielles de ce groupe nous pourrions également utiliser les rayonnements les moins pénétrants. Le graphique nous montre que non seulement les rayons roentgen de 10 et 25 kV, mais aussi les rayons β du radium et du P_{32} pourraient être utilisés.

Matériel clinique des groupes 2 et 3

Ici nous devons aussi combattre l'extension en profondeur. Il faut choisir entre le contact à longue distance (4 cm) et le radium soit sous forme de moulage, soit interstitiel.

Nombre de facteurs jouent ici un rôle. Les doses de radium que nous employons sont, si on les compare à celles de la littérature (4, 6, 7, 8) plutôt élevées, notamment 3,000 r à la surface, généralement en 24 heures à une distance de 1 cm. Il en résulte souvent une dermatite humide avec ulcération que nous voyons guérir entièrement à la suite d'un traitement approprié (onguent chlorophyllé à 1 %) en quelques semaines. Un mois plus tard, l'angiome est devenu beaucoup plus plat et montre des taches blanchâtres. L'extension dans la profondeur n'est plus active. Trois à quatre mois après le traitement, la cicatrice n'est presque plus déplaisante. Par contre, la dermatite humide occasionnée par une irradiation au moyen de rayons roentgen a des suites beaucoup moins

Fig. 3. Avant le traitement. 16 juin 1949.

Fig. 4. Après le traitement. 14 oct. 1949.

favorables et aboutit à un résultat beaucoup moins esthétique. Cette différence est bien frappante.

Il serait bon de fractionner également l'application de radium, par ex. 2 fois 1,000 r à 6 semaines d'intervalle (LABORDE). Cependant nous n'avons aucune expérience de cette méthode appliquée à des formes sous-cutanées profondes, mais nous l'avons utilisée pour les formes superficielles. Nous avons traité la majorité des cas du groupe 2 au moyen de contact à 4 cm de distance. Les doses variaient entre 510 r et 660 r par séance, le nombre de séances était de 2 à 3, espacées de 1 mois à 1 1/2 mois. Cette méthode est très simple et peut également être appliquée avec exactitude pour des localisations difficiles. Cependant si l'extension en profondeur dépasse 1/2 cm ou si la surface est plus grande que celle du plus grand applicateur, il faut lui préférer le radium. Des dosages trop élevés à la limite de champs multiples qui chevauchent partiellement peuvent influencer défavorablement la cicatrice.

Il est utile de diminuer l'épaisseur de l'angiome durant le traitement au moyen d'une compression locale. Ceci peut se faire tant pour les rayons roentgen que pour le traitement par le radium.

Groupe 4

Ici nous avons à faire à des angiomes qui dépassent les possibilités de notre traitement par le radium et par le contact. Nous ne pouvons obtenir un dosage suffisant dans la profondeur comme pourcentage d'une dose non nuisible à la surface que par l'emploi d'un rayonnement de kilovoltage plus élevé. Nous employons ici des rayons de 100 et de 140 kV et un filtre de 3 mm Al. Les doses sont de 1 à 3 fois 400 r, espacées de 1 à 3 mois. Chez des malades jeunes les résultats de cette méthode peuvent être surprenants (voir fig. 3 et 4).

Malheureusement ce groupe comprend également de malades plus âgés ou même adultes. La résistance est accrue au point de rendre necessaires des doses de 5,000 à 6,000 r si l'on vise à une disparition complète de l'angiome. (Les angiomes semblent perdre avec l'âge leur caractère embryonnaire: dès l'âge d'un an, la diminution de sensibilité se fait déjà remarquer.) Dans nombre de cas il vaut mieux se contenter de résultats partiels. Au moyen de radium interstitiel à des doses de 5,000 r réparties d'une façon aussi homogène que possible dans le territoire entre les aiguilles, nous parvenons à réduire considérablement des tumeurs gênantes.

Nous avons obtenus dans quelques cas des résultats remarquables, par exemple pour des hémangiomes de la langue, de la joue et des lèvres. Un rayonnement homogène traversant toute la lésion mais d'un dosage moins élevé, par exemple de 2,500 r, demeure sans résultats et ne donne pas d'amélioration. Nous avons vu une exception: un petit malade de 5 ans souffrant d'un angiolymphangiome grand comme le poing. Cette tumeur devint nettement plus petite après une dose de 2,000 r avec 180 kV, $^1/_2$ mm Cu: il est possible qu'il s'agissait d'un angiome exceptionnellement radiosensible.

RÉSUMÉ

Les différentes formes d'angiomes sont discutées et classées. Les différentes méthodes de traitement radiologique sont décrites ainsi que les modalités de rayonnement. Un traitement schématique doit être rejetté et chaque angiome considéré séparément.

SUMMARY

The different kinds of angioma are discussed and classified. Radiologic methods of treatment and the different therapeutic ray qualities are dealt with; schematic treatment is disproved of, each angioma having to be judged in its own.

ZUSAMMENFASSUNG

Es werden verschiedene Arten von Angiomen besprochen und klassifiziert. Die radiologischen Behandlungsmethoden und die verschiedenen therapeutischen Strahlenqualitäten werden auseinandergesetzt. Jede schematische Behandlung ist abzuweisen; jedes Angiom muss separat beurteilt werden.

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FROM THE ROENTGEN INSTITUTE OF OSLO TRYGDEKASSE, OSLO, NORWAY

SPONDYLOLYSIS — SPONDYLOLISTHESIS[1]

by

Erling Kierulf

Some confusion seems to exist concerning the terms spondylolysis and spondylolisthesis. In a great number of publications luxation of the intervertebral joints and fracture of the neural arch have been termed spondylolisthesis (cit.: SCHMORL and JUNGHANS).

Spondylolysis is defined as a defect in the interarticular portion of the neural arch. BAKKE points out that a defect in this situation must be termed a spina bifida lateralis as long as a definite lysis is not present. The term spondylolisthesis as used by KILIAN is defined by NEUGEBAUER as an anterior displacement of a vertebral body and its superior articular processes in relation to the inferior articular processes and the spinal process. In general »olisthesis» occurs only in the presence of a defect in the interarticular portions of the neural arch.

CHIARI contends that spondylolisthesis is due to anomalies of the articular processes, while PALMER considers it to be due to rupture of the lumbosacral ligaments. KLEINHAUS and WARD state that the defect does not involve the interarticular portion of the neural arch, but only the so-called arch epiphysis, *i. e.*, the portion between the vertebral body and the neural arch. In his diagram of neural arch defects WILLIS has also indicated a defect between the body and the arch. That the latter lesion actually may occur has been shown by the anatomist HAMMERBECK.

NEUGEBAUER's opinion that the cause of spondylolisthesis is a defect in the interarticular portions of the neural arch is strongly supported by BATES, FRIBERG, KLEINBERG and BURMAN, GALLUCCIO, BAILEY, MESHAN and GARLAND.

The etiology of the disease, however, is still a much disputed problem. Most authors believe in a congenital origin *i. e.* a spondylolysis interarticularis congenita (NEUGEBAUER), while others favour a traumatic

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origin and consider the defect to be a result of altered mechanical conditions with increased strain on the interarticular space.

The anatomist HAMMERBECK writes: »Als Entstehungsursache der Spondylolyse kommt entweder eine doppelte Knochenkernanlage oder eine Knochenkernspaltung während der Embryonalentwicklung in Frage. Eine Entstehung der Fugen durch 'Umbau' im Sinne von LOOSER und MEYER-BURGDORFF ist als unwahrscheinlich abzulehnen. Umbauzonen (im Sinne von Looser) hat bisher noch niemand anatomisch am Zwischengelenkstück gesehen. Eine intrauterin entstandene Spondylolisthesis hat BLUM bei einem 40 cm langen Fötus gefunden.»

In favour of a congenital origin BAILEY points out that the defect in the neural arch may involve more than one segment; the defect is unilateral in 25 % of the cases, and familial cases have been observed. As regards the effect of trauma, great disagreement obviously exists. Some authors attach importance to traumatic factors and postulate that the congenital anomaly remains asymptomatic if no injury be superadded. Still others definitely deny the existence of traumatic spondylolisthesis.

The incidence of spondylolysis and spondylolisthesis is estimated by most authors to be 5 per cent (cit.: FRIBERG). In view of this comparatively frequent occurrence of the disease it is strange that it has not yet been established whether or not a transition from spondylolysis to spondylolisthesis may occur; and if so, why the transition occurs.

Some authors make categoric statements about this problem. BROFELDT (cited from FRIBERG) has stressed the fact that not a single case has been published in which transition from spondylolysis to olisthesis has been proved.

FRIBERG writes: »In the literature published after 1925 representing the radiologic era in the research on spondylolisthesis, there is not a single convincing picture showing a spontaneous change in the position of the presacral vertebra.»

On the basis of personal studies he also writes: »The conclusion must be drawn that there is no slipping of either the spondylolytic or the olisthetic vertebrae during adult life.»

Contradicting this view MESHAN writes: »In summary, whether or not the original defect is developmental or traumatic, there is evidence that trauma, even of minor degrees, may widen the defect, initiate symptoms and even start the displacement.»

These conditions merit attention from the point of view of the significance of repeated minor trauma or a single severe injury as a cause of the sliding forward.

The following case seems to demonstrate a transition from spondylolysis to spondylolisthesis.

Fig. 1. 1939 Spondylolysis.

Fig. 2. 1950 Spondylolisthesis.

When examined in 1939 the patient, a heavy worker who was at that time 30 years old and troubled by low back pain, presented roentgenographic evidence of a spondylolysis (fig. 1).

Since the examination in 1939 he had constantly suffered from pain in the back, and the pain had increased.

In 1948 the patient jumped from a height of about 3 metres. Intense pain in the heels developed shortly afterwards, and this was so severe that a roentgenographic examination was carried out with a view to a possible fracture of the calcaneus. This was not present. No particular aggravation of the pain in the back associated with the trauma in 1948 was noted.

Roentgenographic examination in 1950 (fig. 2) showed spondylolisthesis.

It should be noted that the same focus—film distance has not been used for the exposure of the two roentgenograms. There is possibly a slight increase in the lumbal lordosis in 1950, which might be caused by the sliding forward of the vertebral body. Moreover there is a distinct decrease in height of the intervertebral disc in 1950 as compared to 1939. There is also sclerosis of the adjacent corpora and osteophyte formation on the lowermost vertebra.

The narrowing of the intervertebral space is due to degeneration of the disc, which is to be expected in one so exposed. It seems unlikely

to the author that a decrease in the height of the disc should result in a sliding forward of the upper vertebra if the posterior archs were normal. A retroposition of the corpus may be expected as a result of the oblique position of the articular facets in the intervertebral joints.

Since no particular aggravation of the pain in the back associated with the trauma in 1948 was noted, it could not be determined whether the olisthesis was caused by this injury. The effect of repeated minor trauma in his occupation as a heavy worker cannot be excluded.

SUMMARY

A report is submitted of a case of spondylolysis in adult life with transition into olisthesis, possibly of traumatic origin.

ZUSAMMENFASSUNG

Es wird ein Bericht gegeben über einen Fall von Spondylolysis bei einem Erwachsenen mit Übergang zu Olisthesis, möglicherweise auf traumatischer Grundlage.

RÉSUMÉ

Une note concernant un cas de spondylolyse chez un adulte avec transition vers l'olythèse; l'origine traumatique est envisagée.

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